SELF ASSESSMENT ANSWERS

An interesting case of small bowel obstruction

Q1: What is the diagnosis?

This is a case of mechanical small bowel obstruction secondary to an enterolith/bezoar the likely source of which is jejunal diverticulosis

Q2 : What is the differential diagnosis?

This includes the various intraluminal causes of small bowel obstruction such as:

- True foreign bodies: metallic, plastic.
- Food bolus.
- Gallstones.
- Concretions.

Q3: What are the other possible complications of the primary disease of the small bowel?

The possible complications include:

- Diverticulitis
- Haemorrhage
- Obstruction: 1. True obstruction due to enterolith, diverticulitis, adhesions associated with inflammation, volvulus about the adhesions, and intussuception. 2. Pseudo-obstruction or motility disorder.
- Malabsorption due to stasis and bacterial overgrowth.
- Fistulae.
- Asymptomatic pneumoperitoneum.
- Malignant tumours.

Q4: What are the possible treatment options?

The simplest surgical option for small or crushable enteroliths is to milk them distally into the caecum and allow them to pass naturally. If this is not possible, the treatment of this condition is an enterotomy to remove the enterolith with or without resection of the segment of small bowel involved with diverticulosis. Resection may be advocated if the diverticulosis is localised, and is recommended for the other forms of obstruction, haemorrhage, and patients with malabsorption who do not respond to conservative management.

Discussion

The findings at laparotomy were as follows: dilated loops of small bowel seen to mid-ileum. Obstruction at this point was due to an enterolith with collapsed distal bowel (see fig 3 in questions (p 626) and fig 1 below). Two large jejunal diverticulae 12 and 24 inches from the duodenojejunal flexure seen which were palpably empty. The gallbladder was normal with no palpable gallstones. The enterolith was milked proximally and removed via a longitudinal antimesenteric enterotomy which was closed transversely, without resection of the diverticular segment. At laparotomy it is essential to rule out the other causes of enteroliths such as gallstones (as evidenced by a cholecyst-duodenal/jejunal fistula) and to carefully palpate the entire length of the small bowel including the diverticulae for further enteroliths.

Bezoars are masses of solidified organic or non-biological material commonly found in the stomach or small bowel. Four types have been described based on their composition: phytobezoars (containing fibre and cellulose), trichobezoar, lactobezoars, and miscellaneous. The last group includes medications (hydroscopic bulk laxatives, cholestyramine, non-absorbable antacids, vitamin C tablets, and Isocal tube feeds), parasites (*Ascaris lumbricoides* or roundworm), and synthetic fibre.[1–3] A case of carpet fibre bezoar forming at the site of a stapled intestinal anastamosis in a child with pica has been described.[3] In general, the formation of bezoars in the small intestine appears to be at sites of stasis such as blind loops, tumours, and diverticulae (duodenal, jejunal, and Meckels').[4–7]

The incidence of acquired jejunal diverticulosis varies from 0.2% to 1.3% on necropsy studies to 2.3% on enteroclysis.[8] It is associated in 33% to 75% of cases with diverticula elsewhere in the gastrointestinal tract. Enteroliths that form in the proximal small bowel contain bile salts and are frequently radiolucent whereas as many as a third of those that form in the ileum are radio-opaque because of precipitation of mineral salts in an alkaline environment.[9]

The diagnosis is therefore rarely made on the preoperative plain abdominal radiograph. Computed tomography is the modality of choice for investigating patients with higher grades of small bowel obstruction where early surgical intervention is contemplated.[10] There is an increasing tendency to utilise computed tomography to help define the cause, severity, and complications of small bowel obstruction due to the unreliability of clinical signs to predict accurately those patients requiring early intervention.[11]

This is an unusual cause of small bowel obstruction that needs prompt diagnosis and operative treatment.

Final diagnosis

Enterolith causing small bowel obstruction.

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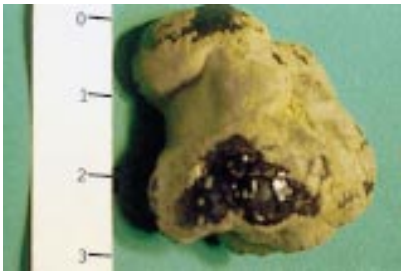

Figure 1 Enterolith.

Iatrogenic groin pain

Q1: What is the likely clinical diagnosis?

The triad of groin pain, hip flexion, and femoral neuropathy indicates iliopsoas sheath haemorrhage. This condition occurs in patients with inherited coagulation disorders, particularly haemophilia A, and in patients on oral anticoagulants.[1 2] Spontaneous haemorrhage occurs deep to the iliacus fascia from the iliacus or psoas muscles, blood tracking from the retroperitoneal space into the pelvic extraperitoneal space. Occasionally massive bleeding can lead to signs of volume deficit.

The iliacus fascia invests the psoas major and iliacus muscles and continues inferiorly as the posterior wall of the femoral sheath. This explains the association with femoral neuropathy, the nerve lying in the groove between the iliacus and psoas muscles. The predilection for the iliacus muscle is unclear.

Q2: What lesion is shown on the computed tomograms (see p 627)?

The computed tomograms shows a collection behind the left iliacus muscle which displaces this anteriorly and separates it from the iliac blade. The left iliopsoas muscle appears enlarged with heterogeneous attenuation internally.

Q3: How should this condition be managed?

In our patient, warfarin was temporarily stopped. He was administered vitamin K, and thereafter started on heparin. The international normalised ratio came down from 7.2 to 2.0 within 24 hours. The pain resolved. There was some residual non-disabling thigh weakness at the time of discharge.

Discussion

Haemorrhage into the iliacus and or psoas muscles is a well recognised complication of overanticoagulation, as well as of haemophiliac disorders. The precise incidence and initiating mechanism of this condition is unclear. Two anatomical syndromes have been described.

Spontaneous haemorrhage may commence either in the iliacus muscle, in which case bleeding occurs deep to the iliacus fascia and a femoral neuropathy may coexist. Alternatively bleeding may commence in the psoas major muscle initially or spread from the iliacus muscle to the psoas. In this case involvement of other components of the lumbosacral plexus, including the obturator nerve and the lateral femoral cutaneous nerve of the

thigh, is likely. A similar clinical picture may be produced by neoplastic infiltration of the lumbosacral plexus.

Pain is the presenting feature, involving the groin, and radiating to the thigh and leg. This is followed by gradually increasing paraesthesiae and limb weakness. A flexion and lateral rotation deformity of the hip may ensue. Passive hip extension is restricted and painful. Delayed development of bruising in the groin may occur. The pain may resolve in a week, with slower and often incomplete recovery of neurological function. In 10%–15% of cases there may be no significant improvement.[3]

There is little definitive guidance on management, as the literature is largely anecdotal and based on case reports or small case series. Overanticoagulation needs to be recognised and corrected. Computed tomography or ultrasound guided aspiration may be helpful, especially if sepsis is suspected.[4]

The prognosis must remain guarded, as residual neurological sequelae are possible even where surgical treatment has been undertaken. With the increasing usage of therapeutic anticoagulation, doctors dealing with anticoagulated patients need to be aware of this clinical presentation.

Final diagnosis

Iliopsoas sheath haemorrhage.

Acknowledgement

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Terminal ileal stricture

Q1: What does the small bowel enema show (see p 627)?

The small bowel enema shows normal jejunum. The ileum is shortened in its distal portion and uniformly narrowed with a smooth outline; the ileocaecal junction is well delineated and the caecum is normal.

Q2: What is the differential diagnosis?

The differential diagnosis of ileal stricture includes tuberculosis, Crohn's disease, pelvic inflammation, ischaemia, radiation enteritis, carcinoid infiltration, lymphoma, and diffuse enteropathy—that is, disorders where there is inflammation, infiltration, or oedema of the small bowel. History and clinical findings in this case did not contribute to the diagnosis.

Q3: How can you confirm the diagnosis?

Enteroscopy/terminal ileoscopy is the investigation of choice. In active Crohn's disease, the terminal ileum shows patchy asymmetrical and heterogenous mucosal lesions. Ulcers which may be aphthoid, superficial, or deep are seen surrounded by normal mucosa. Tuberculoid granuloma is the most specific finding on histology apart from infiltration of lamina propria by lymphocytes and plasma cells with aggretates of lymphocytes near the base of the crypts. In the present case, the smooth and featureless mucosa, and inflammatory cellular infiltrate of lamina propria suggests Crohn's disease in remission. The patient has not been on any medication for over four years.

Discussion

Segmental areas of luminal narrowing of ileum referred to as ileal stricture is due to rigid thickening and fibrosis of its wall resulting in obstruction. It is a common complication of Crohn's disease, tuberculosis, and intestinal ischaemia. A flare-up of inflammatory process causes temporary intestinal narrowing; when healing occurs with a scar or fibrous tissue formation the obstruction is complete. The narrowing could either be circumferential and concentric or eccenteric and irregular in nature. On barium contrast examination, these strictures typically appear as segmental narrowing without normal mucosal pattern and with smooth tapered ends—referred to as the "string sign". Strictures themselves are painless and may not require treatment. But sometimes, these areas become so narrow and result in a partial or total obstruction.

What is the pathophysiology of stricture formation? The intestine can propel the lumenal contents only when the lumen remains fairly wide enough. When there is damage to the intestinal mucosa due to inflammation, the smooth muscle cells of the intestine activate a complex chain of events, involving a host of immune system components, for example, interleukin-1β. There is production and deposition of more than normal collagen at the site of injury. Scarring occurs, the layers of intestinal muscle thicken, and the muscles no longer move smoothly and easily. In short, a stricture forms, compromising the intestine's ability to function efficiently.

An important differential diagnosis of ileal stricture in the present case is Crohn's disease, based on the peroperative findings of mesenteric thickening and ileal stricture at enteroclysis. A featureless outline of a diseased ileal segment, due to atrophy of the folds from long standing inflammation, is not an uncommon finding in Crohn's disease.[1] In one series this was seen in 29% of cases[2]; the biopsy is not likely to be helpful in these situations. Crohn's disease is being increasingly reported from India.[3] It is today included as an important differential diagnosis for ileal tuberculosis. The strictures are caused by shrinkage of a tuberculous ileocaecal mass to form a fibrous constriction. In a country where both problems exist, distinction becomes difficult. Non-response to antituberculosis treatment favours the diagnosis of Crohn's disease. The patient has not been treated for tuberculosis.

In Crohn's disease, like tuberculosis, the small intestine is the most common affected site (80%).[1] In the early stages of the disease, the narrowing is due to oedema and spasm; with progression of the disease, fibrosis manifests as a lumenal narrowing. These findings are also seen in tuberculosis. Few radiological signs are specific for Crohn's disease. These include fissures, ulcers, sinuses, fistulae, and asymmetrical involvement, skip lesions, and long longitudinal ulcers. Less specific findings include luminal narrowing, stricture formation, and dilatation proximal to stenosis, thickening of the mucosal folds, cobblestoning, discrete ulcers, or mural thickening. Long segmental narrowing of the terminal ileum was the only positive finding in the present case.

Khwaja and Subbuswamy reported ischaemic strictures of the small intestine from northern Nigeria.[4] The radiological features are non-specific and simulate tuberculosis and Crohn's disease. Even at laparotomy, it may be difficult to differentiate this from Crohn's disease and tuberculosis. Hypotensive drugs can occasionally produce intestinal ulceration and stricture formation.[5]

The barium infusion technique (enteroclysis) is an ideal investigation for study of the small intestine, both for focal lesions and for extensive mucosal disease. In the best of hands, the procedure gives an optimal radiological-gross pathological correlation and satisfactory evaluation of the extent, depth of the disease, and complications.[6]

A histological difference between Crohn's disease and tuberculosis is not always possible. Supportive information only helps in making a diagnosis. In the case reported, the peroperative findings of ileal stricture resembled Crohn's disease and ileal tuberculosis; terminal ileoscopy and histology was not helpful. The possible diagnosis is Crohn's disease in "remission".

The patient under study has been asymptomatic for four years and is not on any medication. Intervention in ileal strictures is necessary when an individual is symptomatic. Steroids, aminosalicylic acid preparations, immunomodulators, and verapamil have been used during the inflammatory phase; the latter inhibits the smooth muscle cells' response to intestinal injury and prevents collagen deposition. When the affected segment is fibrosed and scarred, endoscopic balloon dilatation can relieve the obstruction. Surgical options include stricturoplasty and resection of the affected segment.

Final diagnosis

Crohn's disease in "remission".

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Lung nodules in a silver polisher

Q1: What do the chest radiograph and HRCT of the lung show (see p 628)?

The chest radiograph shows nodular opacities, predominantly in the upper lobes. The HRCT lung images show diffusely distributed centrilobular nodules without any evidence of fibrosis.

Q2: What is the differential diagnosis of the HRCT scan appearance and the likely diagnosis in this case?

The differential diagnosis of centrilobular nodules with diffuse distribution on HRCT

Box 1: Differential diagnosis of centrilobular nodules on HRCT lung scan

- Infectious bronchiolitis including tuberculosis.
- Pneumoconiosis (coal worker's pneumoconiosis and siderosis).
- Diffuse panbronchiolitis.
- Vasculitis and vascular metastases.
- Sarcoidosis.
- Respiratory bronchiolitis-interstitial lung disease.
- Hypersensitivity pneumonitis

lung scan (see p 628) are infectious bronchiolitis including tuberculosis, pneumoconiosis (coal worker's pneumoconiosis and siderosis), vasculitis and vascular metastases, sarcoidosis, hypersensitivity pneumonitis, diffuse panbronchiolitis, and respiratory bronchiolitis-interstitial lung disease (box 1). The diagnosis in this case was siderosis in view of the clinical presentation, occupational history of prolonged exposure to iron oxide during silver polishing, and characteristic appearance of centrilobular nodules on HRCT of the lungs.

Discussion

Siderosis (synonyms: welder's lung, buffer's lung, or silver polisher's lung) is a non-fibrogenic or a "benign" form of pneumoconiosis due to the inhalation of iron particles. Iron dust is an inorganic, inert, mineral dust with high radiodensity, which neither causes substantial proliferation of reticulin fibres nor gives rise to collagenous fibrosis when retained in the lungs.[1] Occupations leading to siderosis involve exposure to iron oxide dust or fumes and include silver and steel polishing, iron and steel rolling, steel grinding, electric arc welding, fettling, stripping and dressing castings in iron foundries, boiler scaling, and mining iron ores.[1]

Patients are usually asymptomatic unless there is concurrent smoking or contamination of air with other chemicals such as silica or asbestos.[2] They may have a reddish coloured sputum due to exposure to these dusts.[1] Siderosis is therefore essentially a "radiological disorder", due to the presence of very radiodense opacities, but with no functional impairment.[2 3] Iron oxide exposure may be carcinogenic for the human lung.[4 5] The emission of polycyclic aromatic hydrocarbons as pyrolysis products of organic materials used may be responsible, but requires further confirmation.

The pathology of siderosis is characterised by perivascular and peribronchiolar aggregation of dark pigmented iron oxide particles

Box 2: International Labor Office classification of radiographs of pneumoconiosis[7]

- Round opacities are classified according to size as: p, q, or r (p, up to 1.5 mm in diameter; q, 1.5 ~ 3 mm; r, 3 ~ 10 mm).
- Irregular opacities are classified as: s, t, and u (fine, medium, or coarse respectively).
- Combination of round and irregular as: x, y, and z.

Learning points

- Siderosis, baritosis, and stannosis are types of benign pneumoconiosis with radiographic dense opacities.
- Siderosis, also called welder's lung, buffer's lung, or silver polisher's lung is the most common type.
- Usually asymptomatic, respiratory symptoms may be present in smokers or if there is concurrent exposure to silica or asbestos.
- Centrilobular nodules with diffuse distribution are seen on a HRCT lung scan.
- Diagnosis is made on radiological features and occupational exposure.

present extracellularly in alveolar spaces and walls as well as in macrophages.[1] Slight reticulin proliferation may be present in siderosis, but there is no collagenous fibrosis. If fibrosis is present, it is secondary to the presence of crystalline silica.

Radiologically, siderosis presents as centrilobular opacities on HRCT with a uniform distribution throughout the lung fields with no conglomeration. Centrilobular opacities on HRCT lung can be divided into two types according to size.[6] The larger ones are seen as sharply demarcated, rounded nodules and these correspond to the q and r types of pneumoconiosis (box 2) seen on the chest radiograph.[7] The smaller ones, more frequent in number, are seen as relatively ill defined nodular or branching opacities, a few closely spaced dots, or areas of low attenuation on HRCT and represent radiographic type p pneumoconiosis. The nodules are present diffusely and bilaterally, but with upper lobe and posterior predominance.

The differential diagnosis of centrilobular opacities on HRCT lung includes miliary tuberculosis, pneumoconiosis, metastases, sarcoidosis, hypersensitivity pneumonitis, panbronchiolitis, and respiratory bronchiolitis-interstitial lung disease. Some radiographic features are characteristic of the underlying disorder. In miliary tuberculosis, the size of nodules is relatively uniform throughout the lungs. In haematogenous metastases, the metastatic nodules are usually smooth, well defined, and round in shape. They are usually variable in size and do not show ill defined, fine centrilobular nodular or branching opacities. A classical perilymphatic distribution of the nodules is seen in conditions like silicosis, sarcoidosis, lymphangitic metastases, and amyloidosis.

Patients with siderosis require no treatment and the radiological changes of siderosis may regress after cessation of exposure.[3] The diagnosis of siderosis should be considered in relevant occupations with characteristic radiological abnormalities and absence of respiratory symptoms.

Final diagnosis

Siderosis.

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Spinal tumour with raised intracranial pressure

Q1: Based on history and clinical examination what is the differential diagnosis?

Differential diagnosis for raised intracranial pressure with cauda equina syndrome include entities with lesions at multiple sites such as:

- Malignant meningitis, particularly lymphoma with root lesions.
- Neurofibromatosis.
- Tuberculosis.
- Metastases.

Q2: What are the investigations required?

Imaging of the craniospinal axis and cerebrospinal fluid (CSF) analysis are required for diagnosis. In our case, magnetic resonance imaging of the brain showed no expanding mass lesion, except for a small left parietal arachnoid cyst. CSF analysis was then done by lumbar puncture. The fluid was xanthochromic with an opening pressure of 30 cm H_2O. CSF protein was 30 g/l, glucose 5.4 mmol/l, white blood cell count 40×10^6/l, and red blood cell count 160×10^6/l. This was suggestive of a CSF block in the spinal canal.

Magnetic resonance imaging of the spine showed a intradural, extramedullary heterogeneously enhancing mass lesion at the D11–L2 level, which was hypointense on a T1 weighted image and hyperintense on a T2 weighted image (figs 1 and 2).

Q3: What are the causes of bilateral papilloedema without an expanding intracranial mass lesion?

The causes of bilateral papilloedema without an intracranial expanding mass lesion include:

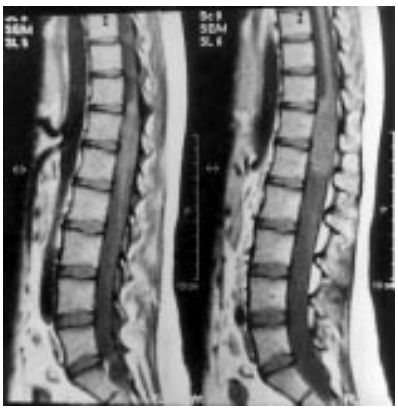

Figure 1 T1 weighted gado enhanced sagittal magnetic resonance image of the thoracolumbar spine showing a well circumscribed 5 × 2 cm mass at D11–L2 level.

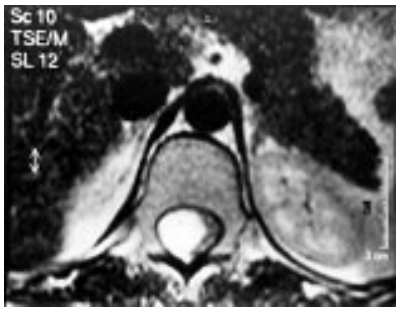

Figure 2 T2 weighted axial magnetic resonance image of the thoracic spine showing a hyperintense intradural mass, more to the right, opposite the D11–L2 vertebrae.

- Hypertension.
- Collagen vascular diseases.
- Guillain-Barré syndrome.[1]
- Idiopathic intracranial hypertension.
- Spinal tumour.[2–4]

Q4: What are the mechanisms/ processes which lead to raised intracranial pressure in patients without intracranial mass lesions?

Bilateral papilloedema and diplopia without an expanding intracranial mass lesion in a patients with spinal cord tumours have been reported in the past. Raised CSF protein (causing delayed absorption of CSF due to increased viscocity) and leptomeningeal inflammation (probably due to the toxic effect of protein secreted by the spinal cord tumours) are the proposed mechanisms for the raised intracranial pressure in patients with spinal cord tumours.

Discussion

This patient underwent excision of the tumour and the biopsy was reported as a myxopapillary ependymoma. After this her raised intracranial pressure and lateral rectus palsy resolved. Bilateral papilloedema and diplopia without an expanding intracranial mass lesion have been reported in patients with spinal cord tumours. Common spinal tumours associated with raised intracranial pressure are ependymoma, schwannoma, meningioma, neurofibroma, and glioma.[3 5] Common clinical features seen among the patients in the previous reported cases include[2–5]:

- Bilateral papilloedema.
- Diplopia.
- Raised CSF protein.
- Young, female patient.

So far no definite cause has been explained for the raised intracranial pressure in these cases. Whether it is because of the tumour itself or because of the effects of the proteins secreted by these tumours into the CSF is not clear.

Raised CSF proteins causing increased viscocity could delay CSF absorption and in turn cause raised intracranial pressure.[6] Clogging of the pores of the semipermeable membrane of the arachnoid villi by large protein molecules could also retard CSF absorption.[7] Raised CSF pressure causes increased pressure on the subarachnoid veins and leads to transudation of substances and further elevation of protein.

Learning point

Consider a spinal tumour in a patient with normal cranial imaging and a diagnosis of presumed benign intracranial pressure.

Venous stasis by the tumour compression of spinal or medullary venous plexuses causing an unfavourable transarachnoid villous hydrostatic pressure is another proposed mechanism.[8] Leptomeningeal inflammation due to the toxic effect of CSF protein causing compromise of CSF absorption is another possible mechanism for raised intracranial pressure.[9]

The spinal canal acts as an elastic reservoir for CSF. With normal changes in physiological cerebral blood flow, the ability of the CSF to flow into and out of the spinal canal is thought to be important in maintenance of a constant intracranial volume. By compromising this system spinal tumours may in fact reduce the capacity of this resorvoir and cause papilloedema.[10]

Final daignosis

Spinal ependymoma with intracranial hypertension.

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A smoker with an apical mass

Q1: Describe the abnormalities shown in figs 2 and 3 (see p 629)

The chest radiograph suggests that the lesion arises in the posterior mediastinum as it is visualised above the clavicle. Figure 2 is a computed tomogram of the thorax that demonstrates a well marginated lesion extending from the neural canal at right T2/T3 level, with significant widening of the neural foramen at this level. There is no evidence of mediastinal lymphadenopathy or lung parenchymal involvement. Figure 3 is a MRI scan that demonstrates a dumbbell shaped neoplasm measuring 40 × 30 mm arising from the right T2/T3 nerve root. It is of high signal intensity on T2-weighted images with a heterogenous area of lower attenuation centrally. The lesion measures 15 mm within the canal and 21 × 30 mm within the thorax and is expanding the neural canal at this level, consistent with a neoplasm of neural origin. The medial aspect of the tumour extends to the lateral border of the spinal cord, which is minimally displaced.

Q2: What is the differential diagnosis?

Mediastinal neoplasms encompass a long list of histologically diverse lesions that can arise from a wide variety of mediastinal structures. In adults, most primary mediastinal neoplasms can be classified in one of four categories: thymus-derived neoplasms, neurogenic tumours, lymphomas, or germ cell neoplasms.[1] The differential diagnosis of masses in the posterior mediastinum includes neurogenic neoplasms (often neurofibromas and schwannomas), aneurysms of the descending aorta, oesophageal tumours, infectious processes including abscesses, and disorders of the thoracic spine, such as sarcomas. In adults, most posterior mediastinal tumours are of nerve sheath origin and are often benign and asymptomatic.

Q3: What was the definitive procedure?

Given the likelihood of this lesion being a benign neurogenic neoplasm, the patient proceeded to a right posterolateral thoracotomy with excision of the lesion.

Q4: Based on the radiological appearances, what potential complication might arise if the lesion was left untreated?

The MRI scan (see p 629) demonstrates that the tumour extends medially to the lateral border of the spinal cord and is causing minimal displacement of the cord. If the tumour were to extend further medially, neurological symptoms resulting from cord compression might arise. Similarly, if the lesion was to further deform the structure of the vertebral body, vertebral collapse might arise.

Outcome

In view of the proximity of the lesion to the spinal cord, thoracotomy rather than thoracoscopic resection was considered the preferred surgical approach. The patient proceeded to a right posterolateral thoracotomy in the fourth intercostal space, and a soft yellow dumbbell shaped mass measuring 30 × 30 mm was discovered in the right third intercostal space, extending into the transverse foramen. The lesion was excised and histological examination showed a partially encapsulated tumour composed of spindle cells in a loose fibrillary background. Antoni type A and B components were present with focal Verocay body formation. The histopathologist concluded that the lesion was a benign intercostal schwannoma. The patient had an uneventful postoperative recovery. Four months after her operation, she remains well, her symptoms have resolved, and there is no radiological evidence of recurrence of the lesion.

Discussion

Neurogenic mediastinal tumours account for approximately 20% of all mediastinal tumours in adults and 40% in children. These tumours may be of nerve sheath, autonomic ganglia, or paraganglionic origin and are generally located in the posterior mediastinum. Neurogenic tumours of the mediastinum with an

intraspinal component connected by a narrowed segment in the intervertebral canal are generally described as dumbbell or hourglass tumours. Tumours of nerve sheath origin classically present as paraspinal masses. These tumours may be neurofibromas, schwannomas, granular cell tumours, melanotic schwannomas, or malignant schwannomas. Schwannomas are most often benign tumours originating from the sheath of peripheral nerves and usually present as solitary and well encapsulated tumours. Most are asymptomatic and are detected incidentally on a routine chest radiograph. Schwannomas arising from the intercostal nerve are well described and may be multiple. Intrathoracic schwannomas may also arise from other nerves, such as the phrenic and glossopharyngeal nerves.[2] Schwannomas are usually benign but may occasionally be malignant. Malignant schwannomas have been described in patients with neurofibromatosis.[3]

Of special concern in the management of neurogenic tumours arising in the thorax is spinal cord compression resulting from either intraspinal lesions or vertebral body destruction and collapse.[4] Predictably, this complication makes for a more difficult surgical excision. Intercostal schwannomas have been successfully resected thoracoscopically. This technique may reduce hospital stay and minimise postoperative complications, but is best reserved for lesions in which there is no evidence of intraspinal extension.[5]

The diagnosis of mediastinal tumours has been aided by recent advances in computed tomography, MRI, ultrasonography, radionucleotide scanning, and fine needle aspiration. MRI is the preferred modality for imaging neurogenic tumours, because of its multiplanar capability and high contrast resolution. MRI can best demonstrate the number and nature of the lesions (differentiating cysts from neoplasms) and the intraspinal and craniocaudad extent of the lesion, and thus determine the optimal surgical approach for patients. This technique is also especially useful for evaluation of the mediastinum of patients for whom the administration of iodinated contrast material is contraindicated. In cases where intraspinal extension is demonstrated, open thoracotomy is the more suitable approach, as the likelihood of conversion to open thoracotomy during thoracoscopic resection is high. However, the definitive diagnosis of schwannoma requires histological examination.

Final diagnosis

Benign schwannoma, fourth intercostal space.

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A man with a chest mass

Q1: What is the interpretation of the biopsy specimen (see p 629)?

The specimen is an excision biopsy of the tumour. Cut section of the mass shows a variegated appearance with areas of haemorrhage, cystic changes, and necrosis. The histological findings are those of metastatic adenocarcinoma.

Q2: What are the different types of rib tumours?

The bone metastases is either osteoclastic (often squamous cell carcinoma), osteolytic (prostate carcinoma, poorly differentiated adenocarcinoma, and breast carcinoma in young patients), intratrabecular, or of mixed type.

Q3: What is the mode of spread to the rib?

Bone marrow metastases is either via the transpulmonary route, or the vertebral venous system—the latter results in metastases to the spine.[1] The frequency of bone metastases via the vertebral venous plexus without pulmonary metastases is 30% for carcinoma of the prostate, 10.4% for uterus, 7.4% for breast, and 3.5% for stomach.[1]

Discussion

The patient has a rib secondary from an obscure primary. Detailed gastrointestinal tract evaluation and chest evaluation were non-contributory.

Moriwaki has described different types of rib tumours.[1] These can be further classified as benign or malignant—the latter are often secondaries from a primary in the lung (35%).[2]

Micrometastases from gastro-oesophageal malignancy has been reported.[1 3] Oesophagogastric malignancy results in micrometastases of ribs in 88% and in the iliac crest in 15%.[3] These are independent of the histological type, nodal status, and neoadjuvant therapy.[3] Macrometastases is less common.

Yang *et al* described three sonographic patterns on cross section of abnormal ribs[4]; ultrasound guided biopsy yields a 100% result without any complications and is an ideal method of confirming the diagnosis.

Management depends on the histopathology and is often directed to the primary site. Large tumours, as in our case, requires surgical resection followed by chemotherapy. The patient is doing well six months after the resection. Failure to find the primary tumour poses problems in management. Yet it remains controversial whether the prognosis improves when the primary tumour is identified by intensive diagnostic search as in the present case.[5] Overall the prognosis is poor.

Final diagnosis

Adenocarcinoma of the rib.

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Terminal ileal stricture

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Notes