Giant pre-sacral neurofibroma

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Summary: We report a case of pre-sacral neurofibroma diagnosed on a routine medical examination. The pathological features and management criteria of these rare tumours are discussed.

Introduction

Neural sheath tumours are a sub-group of soft tissue neoplasms encountered in the cutaneous nerves of extremities, head, neck and trunk. These are occasionally found in the mediastinum and the retroperitoneal tissues of the abdomen. We describe a case of giant retroperitoneal neurofibroma arising from the pre-sacral area in the pelvis. The extreme rarity of this occurrence prompted us to report this case.

Case report

A 52 year old man was found to have a mass in the lower abdomen on a routine medical examination. On questioning he admitted having a poor urinary flow and post-micturition dribbling. Abdominal examination revealed a firm supra-pubic mass extending to the umbilical region. The renal function was within normal limits. Ultrasound scan demonstrated bilateral hydroureteronephrosis, a normal bladder and a large solid mass arising from the pelvic cavity, and extending into the abdominal cavity. Intravenous urography confirmed bilateral ureteric obstruction with hold up of the contrast at the vesicoureteric junctions due to extrinsic compression by the mass; the bladder was pushed antero-superiorly (Figure 1). Computerized tomographic (CT) scan showed a well defined soft tissue pelvic mass with a maximum diameter of 15 cm, containing areas of calcification and necrosis and displacing the bladder anteriorly and above the pelvis (Figure 2).

Figure 1 Intravenous urogram showing bilateral hydroureteronephrosis, lateral displacement of both ureters by the pelvic mass.

Exploratory laparotomy was performed through a midline incision. A large and almost totally intra-abdominal bladder presented on opening the abdomen. There was a large retroperitoneal tumour arising from the front of the sacrum occupying the whole pelvis and extending up the abdomen retroperitoneally to the level of third lumbar vertebra, closely adherent to the aorta, inferior vena cava and the left common iliac artery and vein. The mass was densely attached to the

Figure 2 CT scan showing a mass arising from the pelvis causing compression and distortion of the bladder. T = tumour, B = bladder, S = sacrum.

sacrum; however it could be dissected out completely. Histological examination showed an encapsulated soft tissue tumour containing interlacing bundles of elongated cells with wavy bipolar nuclei. The cells were associated with fine strands of collagen and a moderate amount of mucoid material separated the cells and the collagen. The stroma contained mast cells, lymphocytes, plasma cells and haemosiderin laden macrophages. There was evidence of perivascular hyalinisation, cystification, calcification and haemorrhage. The appearances were of a benign nerve sheath tumour with the histological features of a neurofibroma.

Follow-up at 6 months showed no evidence of recurrent disease. The symptoms of urinary out-flow obstruction had resolved.

References

- Colombeau, P., Thenot, P. & Suberville, M. Anuria and pelvic cancer. *J Urol (Paris)* 1989, **95**: 54–56.
- Sørensen, C., Kristiansen, V.B. & Luke, M. Infravesical obstruction caused by a retrovesical malignant Schwannoma. *Scand J Urol Nephrol* 1987, **21**: 155–157.
- Stout, A.P. The peripheral manifestations of the specific nerve sheath tumour (neurilemoma). *Am J Cancer* 1935, **24**: 751.
- Donnelly, B.A. Primary retroperitoneal tumours, a report of 95 cases and review of the literature. *Surg Gynecol Obstet* 1946, **83**: 705.
- Melicow, M.M. Primary tumours of the retroperitoneum: a clinicopathological analysis of 162 cases: review of the literature and tables of classification. *J Int Coll Surg* 1953, **19**: 401.

Discussion

Pelvic tumours are known to present with symptoms of urinary obstruction. Usually such tumours lead to ureteric obstruction; although occasionally infra-vesical obstruction may be the presenting symptom.^{1–2} Literature survey revealed only one previously reported case of a pelvic neural sheath tumour presenting with symptoms of urinary out-flow obstruction.²

Neural sheath tumours form a very small proportion of primary retroperitoneal neoplasms. Of the combined total of 553 patients with primary retroperitoneal tumours described in 5 major reviews on the subject,^{3–7} only 7 patients were reported to have tumours of neural sheath origin. The largest study pertaining exclusively to retroperitoneal nerve sheath tumours was by Guz *et al.*⁸ Of the 9 patients described in this report only 2 presented with a pelvic mass, and one of these was a neurofibroma.

Nerve sheath tumours are either benign or malignant. The benign variants are Schwannoma (neurilemoma) and neurofibroma. Malignant nerve sheath tumours are usually referred to as malignant Schwannoma. These tumours often produce very few symptoms until they have grown extremely large in the unrestricted retroperitoneal space. This was so in our case since the tumour was detected on a routine medical examination. Our case also supports the findings by other workers that the tumour size neither denotes the biological aggressiveness nor the malignant potential of the tumour. Furthermore, the feasibility of complete excision and cure cannot be judged on the basis of the size of the tumour alone and every attempt ought to be made to excise the tumour completely.

- Pack, G.T. & Tabah, E.J. Collective review of primary retroperitoneal tumours: a study of 120 cases. *Surg Gynecol Obstet* 1954, **99**: 209.
- Braasch, J.W. & Mon, A.B. Primary retroperitoneal tumours. *Surg Clin North Am* 1967, **47**: 663.
- Guz, B.V., Wood, D.P., Montie, J.E. *et al.* Retroperitoneal neural sheath tumours: Cleveland clinic experience. *J Urol* 1989, **142**: 1434–1437.