Necropsy

There was erosion of both frontal bones and sphenoid with firm, haemorrhagic tumour which was well encapsulated. The tumour infiltrated both orbits and into the left maxillary antrum (Fig. 2). There was fibrosis of the pituitary and atrophy of the optic chiasma and nerves. No myeloma deposits were seen elsewhere in the skeleton. Histological examination showed the tumour to be a typical myeloma. Examination of the bone marrow did not reveal any increase in the number of plasma cells.

Discussion

Willis (1960) has stressed the difficulty of being certain that an apparently solitary myeloma is not, in fact, a precocious first lesion of myelomatosis. The disseminated nature of myelomatosis usually becomes manifest within 2 years of the first appearance of a single myeloma. However, there have been several reports of up to 12 years elapsing between initial presentation as a single myeloma and dissemination occurring (Yentis, 1956; Lumb, 1952).

Willis (1960) has suggested that a solitary myeloma may reasonably be presumed if there is: (a) a long history, (b) histological confirmation, (c) extensive radiological surveys, and (d) post-mortem evidence.

This case fulfills most of the criteria for inclusion in the group of solitary myeloma of bone. There was a 3-year history, histological evidence

of the nature of the single tumour and no evidence of any other medullary lesion in the skeleton at autopsy.

Although involvement of the skull is so characteristic of myelomatosis it is of interest that the skull is so rarely involved by a solitary myeloma. A search of the literature has shown three recorded cases of orbital involvement (Jim, 1955; Cogan, 1956; Björnberg, 1962); but none in which both orbits have been invaded by a solitary myeloma.

Summary

A case of solitary myeloma of bone involving both orbits is presented and discussed.

Acknowledgment

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A case of aortic arch syndrome of Takayasu due to disseminated lupus erythematosus

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THE aortic arch syndrome of Takayasu, variously called 'pulseless disease', 'reversed coarctation', Martorell syndrome and branchial arteritis, is found in young people, mostly women. It is characterized by thrombosis of the vessels arising from the arch of the aorta and causes cerebral, ocular and ischaemic symptoms in the upper extremities. Its aetiology and pathogenesis are obscure (Strachan, 1966).

The case of aortic arch syndrome presented below is interesting because the patient sub-

sequently showed evidence of disseminated lupus erythematosus. It is suggested that the aortic arch syndrome might have been due to involvement of the vasa vasorum of the arteries concerned in the course of disseminated lupus erythematosus, and that dissemination occurred after an interval, the initial symptoms and signs being also caused by this connective tissue disorder.

Case report

The patient is a housewife and accounting machine operator. She was first seen in the out-patient department in November 1956, when aged 25 years. She complained that for 6 weeks she had

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experienced severe pain in the upper chest and right shoulder related to coughing and breathing. She had also felt generally unwell and had sweated profusely. She had had no previous illness. She had only smoked an occasional cigarette.

On examination she was thin and looked ill. Her oral temperature was 100.2°F. Both radial pulses were easily palpable and the blood pressure was 140/70 mmHg in the right arm. There was diminished movement of the right lower ribs. The percussion note was impaired and a pleural friction rub was heard in the same area. There were no skin lesions, joint changes or palpable lymph glands. The fundi oculi appeared normal. She was admitted to hospital with a tentative diagnosis of primary tuberculosis. The radial, carotid, temporal, dorsalis pedis and posterior tibial pulses were palpable on both sides at that time.

Investigations: Chest X-ray: some fine discrete mottling in the right lower zone. Hb, 9.6 g/100 ml; blood film, normocytosis; white cell count, 8300/cm³; normal differential; ESR 57 mm/hr (Wintrobe). C reactive protein, strong positive; LE cells were not found on four occasions. Direct Coombs' test, negative in serial dilutions; sternal marrow, no abnormality seen; urine, no proteinuria or glycosuria; no abnormality seen on microscopy of the centrifuged deposit. Blood urea, 24 mg/100 ml; plasma proteins: total, 7.2 g/100 ml; albumin, 4.2 g/100 ml; globulin, 3.0 g/100 ml; electrophoresis, increase in γ and α_2 -globulins; CSF, normal.

Throat swab; no pathogens were isolated; guinea-pig inoculation was negative. Gastric washings, no acid-alcohol fast bacilli seen or cultured. Mantoux skin testing, negative up to a dilution of 1 : 10. Blood cultures, repeatedly sterile; WR, negative; antistreptolysin titre, repeatedly normal; Widal, no rise in titre against *Br. abortus*; positive at a titre of 1 : 60 against *S. typhi* 'O' antigen only. This remained the same on repeated testing and was thought to be an anamnestic reaction associated with a persistent unvarying rise of 1 : 40 in the complement fixation test titre against psittacosis group antigen. Her family had had a sick parrot 10 years previously. Complement fixation tests for influenza A, B, C and D, the APC group and 'Q' fever were all negative. Agglutination tests for streptococcus MG were also negative. No cold agglutins were detected. Complement fixation and dye tests for toxoplasmosis were negative.

During this hospital admission the patient had an intermittent pyrexia rising to 101°F which persisted although there was a partial response to

aspirin 80 gr. daily. It was decided that she should be treated with antituberculous drugs pending the completion of the investigations. She was given isoniazid 100 mg b.d. and streptomycin 1 g b.d. for 1 week and 1 g daily thereafter. The chest lesion cleared clinically and radiologically, but the fever continued and the erythrocyte sedimentation rate remained consistently high. Repeated white cell counts were normal and the haemoglobin concentration did not change appreciably.

In January 1957 she began to complain of dizziness and of pains in both shoulders and arms. Streptomycin was withheld after she had received a total dose of 38 g over a period of 24 days, and *para*-amino salicylic acid was substituted. There were no signs of cerebellar disturbance but her hearing was impaired. The patient was re-admitted to hospital where the ward sister found that her radial pulses were difficult to feel. A few days later they were both impalpable as were the subclavian pulses, while pulsation in the carotids was felt only with difficulty. It was impossible to record the blood pressure in her arms: it was found to be 170/110 mmHg in the legs. The patient now complained that the pains in her arms were related to effort or to raising these limbs. At this time an ophthalmological examination was normal.

The diagnosis was made of aortic arch syndrome.

Chemotherapy was discontinued. Prednisolone 10 mg t.d.s. was substituted and her pyrexia regressed within 48 hr. She has been afebrile since. The dosage was gradually reduced to 5 mg b.d.

The patient was repeatedly observed as an out-patient. Her general condition improved and her fundi remained normal. The subclavian and carotid pulses felt normal by November 1957: both radial pulses were palpable but weak in January 1959 but by March 1960 they too became normal.

On one occasion in 1957 she had a syncopal attack whilst extending her head when washing. In April of the same year plethysmographic studies were undertaken showing results within normal limits. Other investigations were continued as an out-patient. The ESR fell to 15 mm/hr (Wintrobe) in April 1957, plasma proteins were normal in May 1958, but electrophoresis still showed an increase in the globulin fraction. The blood urea remained normal. LE cells were not detected but the anti-nuclear factor was positive in November 1962.

In December 1962 the radial, subclavian and carotid pulses were strong and equal and the blood pressure could be recorded as within normal limits in both arms. However, she still complained of a slight aching in the arms, especially the forearms, which was provoked by excessive use

and exacerbated further if the limbs were elevated at the same time.

A systolic murmur could be heard to the left of the umbilicus, quite localized, over the left renal artery. This suggested the possibility of a renal stenosis as a further development of her arterial disease.

An intravenous pyelogram showed a good renal outline on both sides, the left kidney being 1 cm shorter than the right. An aortic arch and renal angiogram were done. The former showed narrowing of both subclavian arteries from about the level of the first rib and thence distally. There was no major narrowing at the origin of the great vessels and the aorta appeared normal. The renal angiogram was normal.

After these investigations the steroid therapy was gradually reduced further.

Chest X-rays in January 1964 and March 1965 showed no abnormalities: the patient stopped her prednisolone in December 1964 after a course lasting 7 years. It was after this that the patient first developed Raynaud's phenomenon though the pulses were still normal.

In March 1965 the patient developed symmetrical, purplish, erythematous scaly lesions on the lateral and posterior aspects of both legs. Once again no LE cells could be found in the blood. There was no albuminuria and no pyrexia. There was still a slight increase in the γ -globulin fraction of the serum and there was also a raised ESR of 42 mm/hr (Wintrobe). The leucocyte count was only 4000/mm³.

The skin lesions were thought to be of systemic lupus erythematosus and a skin biopsy was taken. This showed a round celled infiltration compatible

with early lupus erythematosus.

The patient was restarted on prednisolone. She has since been well, there has been no recurrence of the vascular disorder, no Raynaud's phenomenon, no skin lesions and no pyrexia.

All the haematological tests which were previously abnormal have now returned to normal.

Comment

There seems no doubt that our patient has disseminated lupus erythematosus. It seems possible that this connective tissue disorder had previously involved the vasa vasorum of the vessels arising from the arch of the aorta, so giving her the signs and symptoms of the aortic arch syndrome.

It seems very unlikely that the disappearance of the pulses could have been due to an occlusion of the vessels concerned as such excellent recanalization would be unusual after such an event.

It is interesting to note that the pulse abnormalities developed whilst the patient was receiving streptomycin. This drug might have been the cause of an exacerbation of the disease process. The skin lesions occurred shortly after stopping steroid therapy. They may have been provoked by this.

It is also noteworthy that this case of aortic arch syndrome of Takayasu seems to be the first that has been observed from its onset and also the first that responded completely to steroid therapy.

Acknowledgment

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