BONE LESIONS SECONDARY TO BENIGN PHAECHROMOCYTOMA

FOUR CASES IN CHILDHOOD

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Metaphyseal sclerotic bone changes associated with benign phaeochromocytoma are very rare in childhood. We report four cases, in each of which the radiographic changes returned to normal after removal of the tumour.

Four children with benign phaeochromocytomas who had hypertension as the main symptom also had bone lesions. The urinary excretion of catecholamines was elevated in each case.

Case 1. A five-year-old girl complained of pain in the right upper limb. Her chest radiograph showed an opacity at the right apex of the right lung and films of the knees showed dense sclerosis of the distal femoral and proximal tibial metaphyses with some circumscribed lucent areas (Fig. 1a). Two weeks after these films were taken, a phaeochromocytoma was removed from the apex of the right lung. One year later the sclerotic areas of the distal femur were less marked but more diffuse with more lucent areas, and the sclerosis of the proximal tibiae had almost disappeared (Fig. 1b). Some 13 months after the onset of the disease, phaeochromocytomas were removed from both adrenal glands and four months after this a recurrence was removed from the left adrenal. The films taken 19 months after the onset show that the sclerotic areas of the distal femora had slightly decreased (Fig. 1c). Two months later, another left adrenal tumour was

removed and the films taken at 31 months from the onset show that the sclerosis has largely disappeared (Fig. 1d).

Case 2. A nine-year-old boy complained for two months of pain in the right ankle and knee. Radiographs showed bilateral increase in density of the metaphyses of the knees (Fig. 2a) and of the ankles (Fig. 2b). There was also increased activity of the affected areas on scintigraphy. Biopsy disclosed nonspecific bone-resorption. A benign phaeochromocytoma of the left adrenal was removed. Several months after operation the blood pressure was normal but the radiographs of the knees and ankles showed that the lesions had decreased, but that there were still some abnormalities.

Case 3. A boy aged 11 years had acrocyanosis with swollen and slightly cyanotic hands. Radiographically, the metaphyses of several phalanges looked irregularly dense (Fig. 3a). At operation, bilateral benign adrenal phaeochromocytomas were removed. His previous hypertension regressed and at long-term follow-up the radiographic appearance of the hands is more normal (Fig. 3b).

Case 4. A teenager who complained of headache had the radiographic appearances shown in Figures 4a, 4b and 4c. Bilateral benign phaeochromocytomas were removed. Five years after surgery, the bone lesions had decreased considerably as shown in Figures 4d, 4e and 4f.

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DISCUSSION

Cases such as we describe are very rare. One previous report concerned a 12-year-old patient with a benign phaeochromocytoma and a history of ankle pain; the radiographic changes were suggestive of bone infarcts of

Fig. 1a

Fig. 1b

Fig. 1c

Fig. 1d

Case 1 – (a) appearance at onset; (b) one year later; (c) 19 months later; (d) 31 months after onset (see text).

Fig. 2a

Fig. 2b

Case 2 – (a) showing the sclerosis at the knees, and (b) at the ankles.

Fig. 3a

Fig. 3b

Case 3 – (a) films before removal of the pheochromocytomas; (b) after.

Fig. 4a

Fig. 4d

Fig. 4b

Fig. 4e

Fig. 4c

Case 4 - Original appearance of (a) humerus (b) wrist and (c) ankle; (d) (e) and (f) show the appearances five years after surgery.

Fig. 4f

the distal metaphyses of the tibiae and disappeared after surgery (Becker, Redisch and Messina 1967). Another was of a six-year-old girl with a large phaeochromocytoma and with sclerotic areas involving the distal ends of both femora and radii, and the proximal ends of the tibiae. A radionuclide scan was normal. Films taken 17 months after removal of the tumour demonstrated a normal appearance of the affected metaphyses (Hernandez et al 1980).

The bone lesions in such cases can be explained by

microcirculatory changes, which in turn can be explained by what is known about adrenaline, noradrenaline and phaeochromocytomas. Adrenaline has been shown to cause hypovolemia and haemoconcentration in normal men (Ebert and Stead 1941; Kaltreider, Meneely and Allen 1942). Intravenous infusion of noradrenaline causes an increase in haematocrit entirely due to plasma loss, the total red blood cell mass remaining constant (Finnerty, Buchholz and Guillaudeu 1958). This tendency toward haemoconcentration is responsible for aggrega-

tion of red blood cells, increase in shear stress, and sluggish flow. As the viscosity of the blood increases, the velocity gradient or rate of shear decreases (Wells 1964). These changes lead to engorgement of capillaries and eventually to the formation of microthrombi, resulting in stasis and disruption of the vessel wall.

The changes described are in the surface microcirculation but they may be assumed to be present also in the metaphyses. Stasis of the blood and microthrombi may lead to infarctions such as those seen in the radiographs. The greater vascularity of growing bone means that the changes take place more rapidly in the

child and this explains why such cases have so far been reported only in children.

The radiological differential diagnosis of such dense heterogeneous metaphyses should include the bone infarcts seen in sickle-cell anaemia, chronic symmetrical metaphyseal osteomyelitis (Gustavson and Wilbrand 1974) as well as the metastases of malignant phaeochromocytoma.

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