A child with multiple bony swellings

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A 5-year-old boy presented with multiple bony swellings around his knees, wrists, shoulders, and rib cage. He was in good general health and there was no history of trauma, but his father was concerned about increasing deformity of the child's knees and the gradual increase in size of the swellings around his wrists, chest and knees. On examination, there were bilateral multiple fixed bony outgrowths from the distal radius, medial side of lower femur, upper end of humerus and costochondral junctions. The child had mild genu valgum on the left side. There was no deformity or swelling of the fingers. Movements at the shoulder, hip and knee were within normal limits and there was no neurovascular problem. The father had similar bony swellings around his knees and shoulder but was not bothered about these because they were not growing and he had no functional problems. Plain X-rays of the knees, wrists and chest of the patient and a photograph of him and his father are shown in figures 1-4.

Figure 1 X-Ray of both knees

Figure 2 X-Ray of both wrists

Figure 3 X-Ray of the chest

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Figure 4 Photograph of the child and his father

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Questions

- 1 What are the radiological and clinical findings?
- 2 What is the diagnosis?
- 3 What are the prognosis and treatment?

Answers

QUESTION 1

The X-ray of the knees (figure 1) shows bony outgrowths from the lower femoral metaphyses mainly on the medial aspect. The wrist X-ray shows similar outgrowths involving the distal metaphyses of radius and ulna in both hands. There is also some shortening of the right third metacarpal with broadening of the distal metaphysis. Chest X-ray reveals bilateral outgrowths from the proximal humeral metaphyses with slight broadening of the anterior ends of the lower ribs on the left side.

The clinical photograph of the child shows left-sided genu valgum with diffuse swelling over the lower thighs. There are multiple bony outgrowths at costochondral junctions. The photograph of the father shows bilateral multiple bony growths around the knees and right shoulder.

QUESTION 2

The clinical presentation and the radiological findings are typical of osteochondromatosis, also known as hereditary multiple exostoses, diaphyseal aclasis, and metaphyseal aclasis. The disease should be differentiated from solitary osteochondroma, myositis ossificans and multiple enchondromatosis, the latter involving mainly the hand bones.

QUESTION 3

Most of the time the lesions are asymptomatic and do not affect the general health of the patient. Disability may be produced if the growth is near the joint, interfering with joint movements or irritating the tendon or muscle belly.

Neurological symptoms may be produced by mechanical pressure, especially in vertebral lesions. Rarely, the osteochondroma may become painful because of fracture of the stalk. Malignant transformation to chondrosarcoma is possible, although rare. Surgical treatment is indicated if the tumour is symptomatic or if there is suspicion of malignancy.

Discussion

Osteochondroma is a benign developmental growth defect involving the metaphyseal area of long bones, producing a bony outgrowth with a cartilage cap. It is the commonest benign bone tumour.¹ Osteochondromatosis has a marked hereditary tendency with autosomal dominant inheritance.² The penetrance is almost 100%

Learning points

- osteochondroma is the commonest benign bone tumour
- multiple osteochondroma (osteochondromatosis) differs from solitary lesions in mode of inheritance and chance of malignant transformation
- surgical excision is required for symptomatic and possibly malignant growths

by the age of 12 years.³ In this case, both the boy and his father had multiple osteochondroma. This disorder is usually detected in early childhood around the age of 8-10 years. Long bones are most commonly involved. The majority of patients are asymptomatic and present with fixed bony lumps around the joints. The tumour grows with the growth of the patient and any increase in size of the lesion after skeletal maturity is suggestive of possible malignant transformation.⁴

Osteochondroma has typical radiographic features. The growth may be sessile or pedunculated. Characteristically, the cortex of the lesion is continuous with the cortex of the host bone. The actual size of the growth is greater than the radiological size because of the cartilage cap over the bony outgrowth. The bone harbouring the tumour may be shortened, causing deformity or limb length discrepancy.^{1,2}

If the tumour is symptomatic it should be excised. Excision is done through the base of the tumour along with the cartilage and the perichondrium. The chances of recurrence are greater if the perichondrium is removed incompletely. The exact figure for malignant transformation of the growth is not known but figures usually quoted are 1% for solitary lesions and 6% for multiple lesions.⁵

The relative frequency of chondrosarcoma is greater in central lesions involving the pelvis, shoulder girdle, or spine.³ Malignant transformation should also be suspected if the lesion suddenly becomes symptomatic or begins to grow rapidly. If there is any suspicion of malignancy, the lesion should be excised.

Final diagnosis

Hereditary multiple exostoses.

Keywords: exostoses; bony swellings; chondrosarcoma; osteochondromatosis

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Diarrhoea, fever, shock and bullous skin lesions after ingestion of raw oysters

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A 38-year-old man presented to the emergency department with a 2-day history of cramping abdominal pain, severe vomiting, diarrhoea, and fever with chills. He reported eating a large quantity of raw oysters at a local restaurant, one day prior to the onset of symptoms. The patient had a significant history of daily alcohol consumption (12 cans of beer and one bottle of wine) for many years. On examination, the patient appeared toxic, with a systolic blood pressure of 90 mmHg, pulse of 116 beats/min, respiration rate of 22 breaths/min and an oral temperature of 39°C. Right basal crackles were present on auscultation of the chest. Cardiovascular examination showed tachycardia with normal heart sounds. Abdominal examination revealed a tense abdomen without evidence of ascites, with diffuse tenderness and hyperactive bowel sounds. The patient's skeletal muscles were tender to palpation and movement. Joint examination revealed no evidence of synovitis. A few hours after admission, multiple 1–2 cm skin lesions were noted predominantly on the truncal area (figure). Over the next 24 hours, these evolved into haemorrhagic bullae with purpuric centres.

Laboratory findings were significant for severe leucopenia (1000 cells/mm³), hypoalbuminaemia (1.7 g/dl), abnormal liver function tests (total bilirubin 5.3 g/dl; lactate dehydrogenase 1544 IU/l; aspartate transaminase 615 IU/l; alanine transaminase 465 IU/l), elevated creatine phosphokinase 14 600 IU/l, prolonged prothrombin time 15.9 s, INR 1.6, and lactacidaemia (4.6 mEq/l). Empiric antibiotics using a third generation cephalosporin and doxycycline were started immediately after blood cultures were drawn. However, within 8 hours of admission the patient became diaphoretic, tachypnoeic, hypoxic, and his mental status deteriorated.

Figure Skin showing early target lesions and bullae over the truncal area

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Questions

- 1 What is the diagnosis and what are the predisposing factors?
- 2 What is the treatment and prognosis?