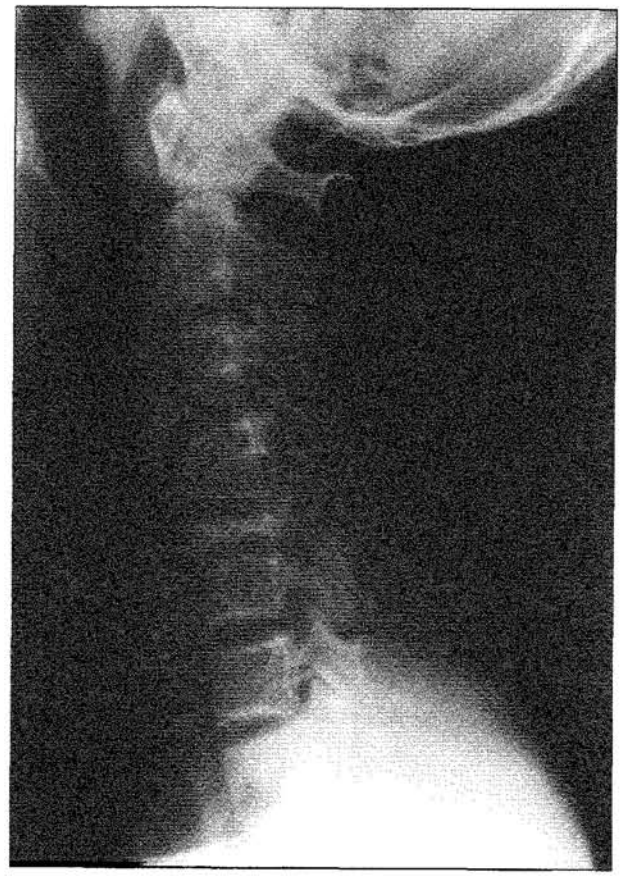
Clinical History:

Incidental radiographic finding (**Figure 1**) in the neck of an asymptomatic 14-year-old boy. He had a minor neck injury 4 years previously and had been thoroughly investigated following that episode of trauma.

Figure 1: Lateral radiograph of the cervical spine



Answer on page 348

What is the diagnosis?

a) Neoplastic disease
b) Infection
c) Langerhans' cell histiocytosis
d) Compression fracture
e) Paget's disease

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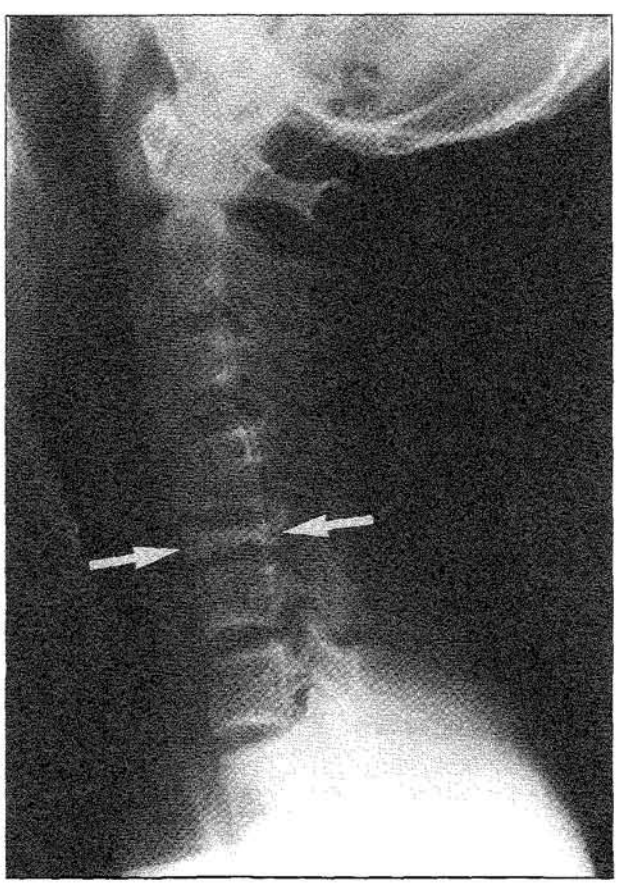
Answer:

d) Langerhans' cell histiocytosis

Radiological findings

The lateral radiograph of the cervical spine (**Figure 2**) shows severe collapse of the fifth cervical (C5) vertebral body. This sign is also known as vertebra plana. The posterior elements are spared. Adjacent disc spaces and vertebral alignment are normal. No pre-vertebral soft tissue swelling is seen. The vertebral bodies at other levels are normal and the bone density is preserved.

Figure 2: Same radiograph as Figure 1. C5 vertebra plana is arrowed. All the cervical vertebral bodies are in alignment, and the intervertebral disc spaces and posterior elements are normal



He had been investigated for neck pain some two months following the previous episode of minor neck trauma. Radiograph performed then showed marked flattening of the C5 vertebral body. As a pathological fracture was suspected, a biopsy was performed which confirmed the diagnosis of eosinophilic granuloma. There has been no significant change in radiographic appearances on follow-up to the present time.

Discussion

Neoplastic disease

Both childhood leukaemia and metastastic neuroblastoma can give rise to vertebral collapse. Leukaemia is the commonest malignancy of childhood. Diffuse medullary invasion often leads to trabecular destruction, osteopaenia and vertebral collapse. In this particular young child, the vertebral collapse is solitary and is not accompanied by osteopaenia, hence the diagnosis of leukaemia is unlikely.[1]

Neuroblastoma is the commonest extracranial solid malignancy in infancy. 75% of the cases occur before the age of 5 years.[2] Skeletal metastasis is common (60%) and can lead to vertebral collapse.[3] History of a known primary tumour is useful for establishing the diagnosis. Besides the clinical history and lack of multiplicity of lesions, the age of this child is too old for this disease.

Infection

Pyogenic and granulomatous infection can produce vertebra plana. The commonest organism responsible for pyogenic infection is staphylococcus aureus.[1] In Asia, tuberculosis is a particularly important differential diagnosis. Infection, whether pyogenic or tuberculous, is often accompanied by destruction of the adjacent disc spaces at the time of presentation. The presence of a paravertebral soft tissue mass makes tuberculous infection more likely.[1] Contiguous vertebral bodies are frequently affected, resulting in at least 2 levels of involvement. The lack of disc and adjacent vertebral

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body involvement in this case makes this diagnosis very unlikely.

Langerhans' cell histiocytosis

This entity was previously known as Histiocytosis X and is an idiopathic, non-neoplastic proliferation of histiocytes that may have either a systemic or localized clinical expression. The disease is classified into eosinophilic granuloma, Hand-Schuller-Christian disease and Letterer-Siwe disease, according to increasing systemic and decreasing skeletal involvement. Eosinophilic granuloma is the localized form and accounts for 60% to 80% of all cases.[4]

Eosinophilic granuloma is most commonly found in children between 4 to 7 years of age. Common presentations are bone pain, local swelling and irritability. It mainly affects the skeleton with solitary (50-70%) or multiple lesions.[2] The long bones, pelvis, skull and flat bones are the commonest sites of involvement. Radiological findings include well-defined medullary lucencies, multilocular lucencies, punched-out lucencies with the classical "beveled edge" appearance in the skull vault, destructive lesions (seen in the skull base, mastoids, sella or mandible) and vertebra plana. It is the most frequent cause of a solitary vertebra plana in childhood. In vertebra plana, the vertebral body becomes wedged and is often wafer-thin. Over time, the vertebral body height is often partially restored.[4] The posterior elements are usually spared and adjacent disc spaces are preserved.[2] Although the thoracic and lumbosacral vertebrae are the usual sites of involvement, the cervical spine can also be affected.

Compression fracture

A combination of flexion and axial loading results in burst fracture of the cervical vertebra. The C5, C6 and C7 levels are typically involved and neurological deficit is common.[5] Burst fractures of the cervical spine characteristically result in loss of vertebral height and fragment retropulsion into the spinal canal.[5] This latter finding was not present in this patient and there was lack of a history of the appropriate mechanism of trauma.

Paget's disease

Paget's disease is a disease affecting middle-aged and elderly patients and is characterized by excessive abnormal bone modelling and bone softening. It is rare among Chinese. The condition is biphasic with an initial active (osteolytic) phase followed by an inactive (osteosclerotic) phase. An intermediate mixed osteolytic – sclerotic phase may also be observed.[1,2] The axial skeleton is predominantly affected. The spine, proximal femur, skull and pelvis are the commonest sites of involvement.[2] In the spine, the lumbar vertebrae are most frequently affected. Typically, the vertebral body is enlarged, with coarsening of trabeculation. Cortical thickening may produce a "picture frame" appearance.[2] Increased density of the vertebral body gives rise to the so-called "ivory vertebra"[1] The softened bone may squeeze out symmetrically in all directions, mimicking collapse due to malignant or infective causes, but the apparently enlarged vertebral body is a helpful differentiating point. The young age of this child and the absence of bony expansion make this diagnosis very unlikely. ■

References

1. Keats TE, Alford BA. Paediatric Musculoskeletal System. In: Grainger RG, Allison DJ. Diagnostic Radiology. 2nd ed. Churchill Livingstone, Edinburgh. 1992;p1739.
2. Chapman S, Nakielny R. *Aids to Radiological Differential Diagnosis*. 3rd ed. W.B. Saunders, London. 1995;pp76-78.
3. Dahnert W. *Radiology Review Manual*. 2nd ed. Williams & Wilkins, Baltimore. 1996;pp676-677.
4. Conway WF, Hayes CW. Miscellaneous lesions of bone. *Radiologic Clinics of North America*. 1993;31:339-358.
5. Resnick D, *Bone and Joint Imaging*. W.B. Saunders, Philadelphia. 1996;p809.