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AN UNUSUAL CASE OF HISTIOCYTOSIS X IN THE SPINE

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A case of spontaneously healing histiocytosis X followed over 9 years is presented. The lesion was located in the cervical and dorsal spine and had unusual radiological features such as soft tissue swelling, involvement of the pedicles and recovery with fusion between adjacent vertebra.

Key words: histiocytosis X; eosinophilic granuloma; vertebra plana; bone tumours

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It has already been shown that skeletal involvement of histiocytosis X can mimic many other conditions (Ennis et al. 1973, Poulsen et al. 1975, Thommesen & Poulsen, unpublished data) and exact diagnosis must in most cases rely on biopsy. However, lesions in the spine in children with development of a vertebra plana are regarded by many as pathognomonic of histiocytosis X until proven otherwise (Ennis et al. 1973) especially if the following radiological features for a solitary lesion are fulfilled:

1. Only one vertebra involved.
2. Adjacent discs intact above and below the diseased vertebra.
3. Disc space about a third wider than the next space above and below.
4. Homogeneous density of the collapsed vertebral body.

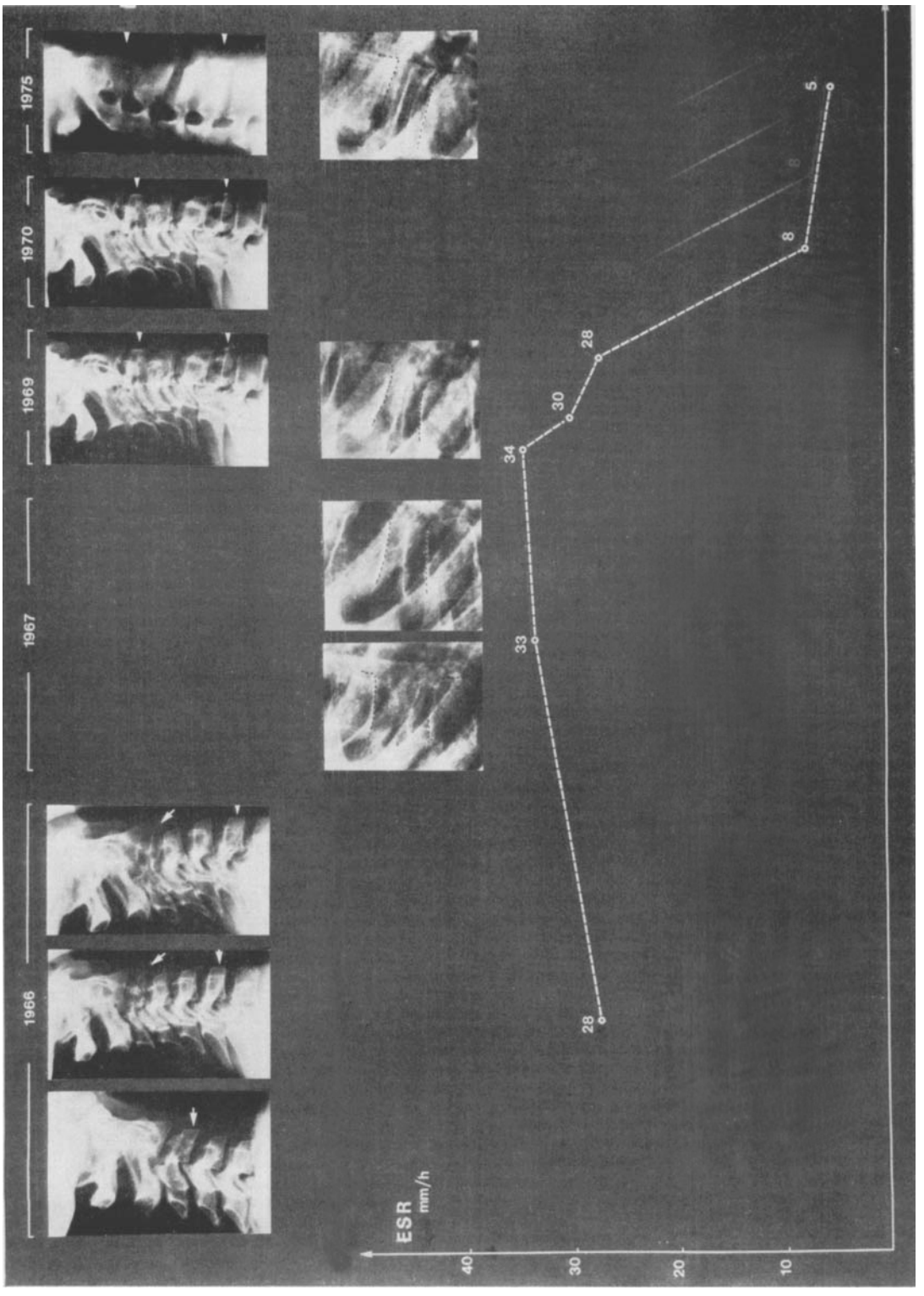
These four classic criteria have been laid down by Compére et al. (1954) in the diagnosis of eosinophilic granuloma. However, the rarity of certain features has been emphasized by Kieffer et al. (1969):

1. Soft tissue swelling is rare.
2. Involvement of the pedicles is rare.
3. Fusion between vertebra after recovery is rare.
4. Cervical involvement is less frequent.

We have recently reviewed a case of probable spontaneously healed histiocytosis X followed over 9 years, which primarily caused some diagnostic problems and during the course showed some less frequent radiological features.

CASE REPORT

An 11-year-old boy was admitted to the Orthopaedic Hospital with pain in the neck after an adequate trauma 1 month previously. The primary X-ray gave rise to a suspicion of subluxation between the third and the fourth vertebral bodies. (Figure 1: first row, first X-ray from 1966). Physical findings were slight limitation of movement in the neck. The patient was treated with traction followed by plaster ban-


1966
1967
1969
1970
1975
ESR
mm/h
40
30
20
10
28
33
34
30
28
8
5

dage. A few months later the diagnosis eosinophilic granuloma was established because of the typical X-ray findings in the cervical spine the same year (Figure 1). Four months later X-ray of the dorsal spine showed partial collapse of the seventh thoracic vertebral body (Figure 1: second row, X-rays from 1967). ESR was 33 mm/h. Serum electrophoresis showed elevated α_2 globulin, normal γ globulin.

Survey X-ray revealed no other lesions. After a short period of local pain he had no symptoms for the next two years, when pain in the right shoulder initiated further X-rays revealing partial collapse of the sixth vertebral body (Figure 1: first row, X-ray from 1969). The X-ray findings were checked and at the last visit in 1975 he was without symptoms and X-ray showed partial regeneration of both the cervical and dorsal lesions.

DISCUSSION

The patient was primarily treated for subluxation between the third and fourth cervical vertebral bodies although the movements of the cervical spine were only slightly limited. Retrospectively one has to acknowledge a slight compression of the third vertebral body with no sign of fracture and a "washed-out" appearance due to osteoporosis. However, the subsequent X-ray picture from 1966 established the diagnosis eosinophilic granuloma based on Compére et al.'s (1954) criteria. It should be noted that soft tissue swelling also is present (Figure 2), and might simulate an inflammatory process, but the ESR level was not characteristic and serum electrophoresis showed a raised α_2 globulin level with no γ globulin response.

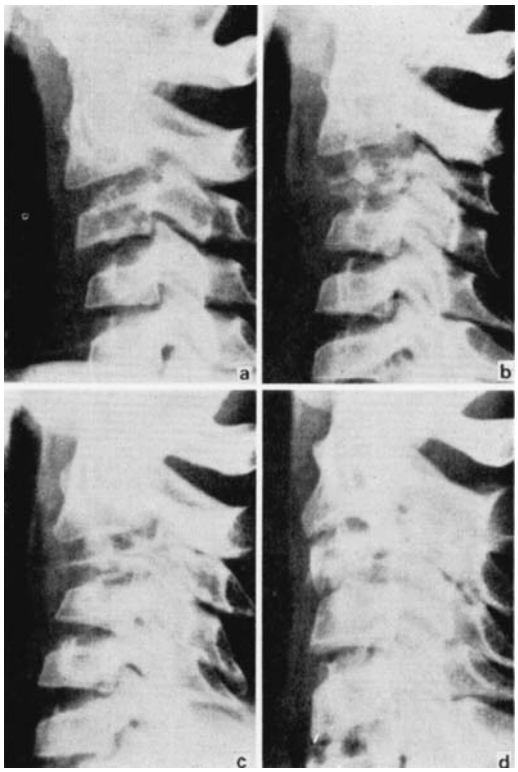


Figure 2. X-rays of the cervical spine over a period of 3 months (a, b, c) illustrate the soft tissue swelling anterior to the third cervical body (b, c); this has disappeared two years later (d).

The course of the affection in the seventh thoracic vertebral body followed the criteria laid down by Compére et al. (1954) with almost complete restoration in 1975 giving rise to the appearance of a "bone within bone" (Figure 1). Comparing the X-rays of the cervical spine from 1966 with those from 1969, it is obvious that also the sixth cervical vertebral body has been involved and recovery has taken place simultaneously with the third vertebral body. Partial fusion with the adjacent vertebra is also seen and furthermore a tomogram from 1975 shows that also the pedicle has been involved resulting in complete fusion

Figure 1. At the top the years of interest to this case presentation are indicated. At the bottom the ESR's in mm/h are shown. The development of the lesions in the cervical spine are shown in the first row of X-ray pictures (cervical segments of interest are indicated by arrows). Note the fusion between the third vertebra and adjacent segments and partial fusion between the sixth and seventh vertebral body, seen on the tomogram in 1975. In the second row of pictures the lesion in the seventh thoracic vertebral body is shown with partial regeneration occurring in 1975.

between those parts in the second, third and fourth cervical segments.

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