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# Infantile Myofibromatosis: A Cause of Vertebra Plana

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**Summary:** We report a case of infantile myofibromatosis causing asymptomatic vertebra plana of L-5.

**Index terms:** Spine, neoplasms; Spine, vertebrae; Children, neoplasms

Infantile myofibromatosis is the most common type of fibromatosis found in childhood (1–3). We present a case of an infant with skeletal and soft-tissue lesions that at biopsy proved to be infantile myofibromatosis. Asymptomatic collapse of the L-5 vertebral body occurred later and was evaluated by magnetic resonance imaging.

## Case Report

A 6-week-old boy was seen in the emergency department with progressive enlargement of head and neck masses. Bilateral periauricular masses had been present since birth and had undergone recent cervical extension. A mass of the right nasolabial fold had been present since 3 weeks of age. The lesions were asymptomatic and the infant was otherwise well. Computed tomography (CT) of the head and neck showed multiple partially calcified masses (Fig 1A and B) that were similar in attenuation to muscle and demonstrated peripheral enhancement after intravenous administration of contrast material. Additional calvarial and metadiaphyseal lytic long-bone lesions were demonstrated on skeletal survey. Some bone lesions had thin zones of marginal sclerosis, whereas margins on others were poorly defined. CT of the chest and abdomen showed no evidence of visceral involvement. Surgical biopsy of one of the neck masses yielded the diagnosis of infantile myofibromatosis.

The patient did well without additional treatment after discharge. Physical examination, plain radiography, and CT examination showed interval enlargement of some masses and diminution of others over the next 5 months. Follow-up skeletal survey at 6 months of age showed enlargement of all of the bone lesions. Features included

better defined sclerotic margins, cortical thinning, and bone expansion. A new lesion was present involving the L-5 vertebral body. A lateral radiograph and magnetic resonance imaging of the spine at 8 months of age showed complete, but asymptomatic, collapse of this vertebra (Fig 2A and B).

## Discussion

Infantile myofibromatosis is the most common type of fibromatosis seen in childhood (1–3). Over the years, it has been described under various synonyms, including congenital generalized fibromatosis, congenital multiple fibromatosis, diffuse congenital fibromatosis, multiple mesenchymal hamartomas, and multiple vascular leiomyomas of the newborn (1). The tumors arise from myofibroblasts that are the precursor cells to both smooth muscle and fibroblasts. Histologically, the tumors show characteristics of both cell lines (1–3). The lesions of infantile myofibromatosis are usually present at birth or are noticed shortly after birth. Almost all patients are seen within the first year of life. Nodules of infantile myofibromatosis can be solitary or diffuse, although solitary nodules are the most common at presentation. Infantile myofibromatosis usually involves the skin, subcutaneous tissues, and bone (1, 2). Visceral involvement is common in the diffuse form and rare in the solitary form. Dural lesions have also been described (4–6).

The prognosis in infantile myofibromatosis is directly related to the site of involvement. Young infants with diffuse infantile myofibromatosis and extensive visceral involvement tend to do poorly, because no definitive treatment is available. Death is usually related to cardiac, pulmonary, or gastrointestinal disease. Isolated

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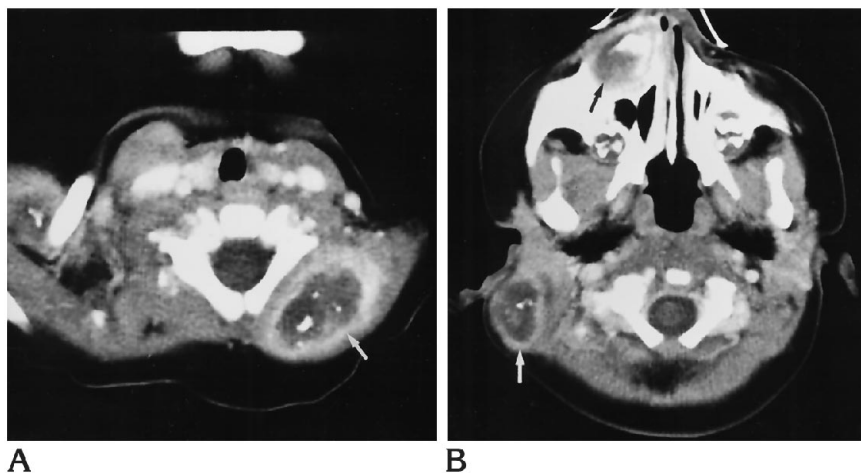


Fig 1. Enhanced axial CT scans at 3 months of age show calcified, inhomogeneously enhancing posterior cervical lesion (arrow, A) and postauricular (white arrow) and nasolabial (black arrow) lesions (B).

skeletal, muscular, and subcutaneous lesions tend to regress over months or years, leaving occasional skeletal deformity (1-3). Treatment usually is conservative, with surgical excision limited to those masses that pose problems because of size or location (3). Often there is an increase in the number and size of the masses before spontaneous regression occurs.

Radiographic features include, as in our patient, soft-tissue masses with frequent calcification and necrosis (1). Lytic lesions with sclerotic borders are often eccentrically located in the long bones (1). In one series, the calvaria, femur, tibia, vertebra, and rib were the most commonly affected bones (1). Asymptomatic vertebral body collapse from spinal involvement with complete regression of lesions and dural involvement with and without adjacent skeletal involvement have also been described (4-8).

The most common cause of vertebra plana in childhood is histiocytosis. The radiographic appearance of vertebra plana is well defined.

There is initially a lytic vertebral body lesion without collapse. Subsequent vertebral compression occurs with partial or complete collapse of the vertebral body. The end plates remain intact, forming the coin- or wafer-shaped vertebra plana. As the growth plates remain intact, there can be subsequent total or nearly total reconstitution of vertebral height. This reconstitution is thought to be age dependent, with younger patients having better recovery of vertebral height (9). Reconstitution of vertebra plana has been described in both infantile myofibromatosis and histiocytosis (7). Less common causes of vertebra plana are lymphoma, leukemia, and metastases. Rare causes include Morquio's disease, Gaucher's disease, hypophosphatasia, and radiotherapy. Although vertebra plana is uncommon in infantile myofibromatosis, it should be considered in the differential diagnosis when an infant has multiple soft-tissue and skeletal lesions.

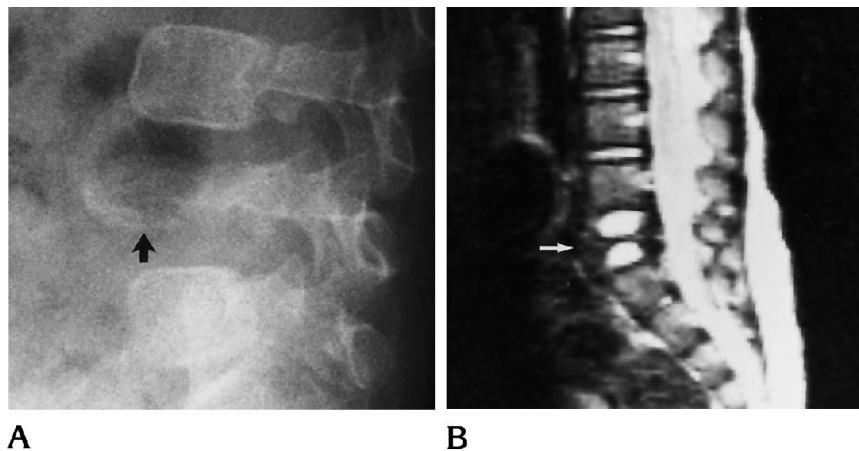


Fig 2. A, Lateral lumbar plain film at 7 months of age shows a lytic lesion (arrow) of the L-5 vertebral body with minimal loss of height.

B, Unenhanced sagittal T2\*-weighted magnetic resonance image (3500/90/1 [repetition time/echo time/excitations]) shows vertebra plana (arrow). Minimal paravertebral soft tissue is seen on the sagittal image.

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