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Juvenile Fibromatosis of the Posterior Mediastinum with Intraspinal Extension

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Summary: Chest radiography, CT, and MR imaging were performed in a 3-year-old girl who had posterior mediastinal fibromatosis with transforaminal intraspinal and chest wall extension. Chest radiographs and CT scans showed a slow-growing, noncalcified but locally aggressive left paravertebral mass. The mass was slightly hyperintense relative to muscle on both T1-weighted and fast spin-echo T2-weighted MR images.

Index terms: Children, neoplasms; Spine, neoplasms

Juvenile fibromatosis is a locally infiltrative pseudoneoplastic process characterized by fibroblastic proliferation (1–4). Fibromatosis may occur anywhere in the body, most commonly in the superficial soft tissues, extremities, or abdomen (2–4). Fibromatosis involving the mediastinum in children is extremely rare (5). We describe a case of juvenile posterior mediastinal fibromatosis with transforaminal intraspinal and chest wall invasion mimicking a paraspinal neuroblastoma.

Case Report

A 3-year-old girl had a 2-week history of a nontender mass on the posterior aspect of the left lower thorax. Physical examination was unremarkable except for a 2 × 2-cm fixed, painless, elastic mass on the left side of the posterior chest wall. No neurologic deficit was noted. The α-fetoprotein, β-human chorionic gonadotropin, and 24-hour vanillylmandelic acid levels were within normal limits. Results of bone scan and bone marrow aspiration were negative.

Chest radiographs revealed a left-sided posterior extrapulmonary mass with erosion of the adjacent 9th and 10th ribs (Fig 1A). A slow-growing posterior mediastinal neurogenic tumor was the initial diagnosis. Computed tomography (CT) showed an enhancing left-sided posterior mediastinal mass with chest wall invasion and possible evidence of intraspinal extension. No intratumoral calcification was detected (Fig 1B). Magnetic resonance (MR) imaging (1.5 T, Signa system, General Electric Medical Systems, Milwaukee, Wis) showed that the mass was slightly hyperintense relative to adjacent muscle on both T1-weighted and fast spin-echo T2-weighted images and that the intravenous administration of gadopentetate dimeglumine enhanced the mass on the MR image. Chest wall invasion and transforaminal extension with extensive intraspinal extradural spread were also seen (Fig 1C and D). Open biopsy of the chest wall mass was done, and microscopic examination revealed fragments of tumor tissue composed of bundles of fibroblasts suggestive of fibromatosis.

At surgery, a 6 × 5 × 3-cm well-defined left-sided posterior mediastinal mass with investment of the adjacent ribs and extension to the back muscles was found. En bloc paraspinous excision of the mass consisted of partial resection of the 9th to 11th ribs, a T-9 to T-10 laminectomy with total removal of the extradural tumor, and reconstruction of the chest wall.

The surgical specimen was a well-demarcated grayish white mass enveloping the ribs in its peripheral part. Microscopic examination revealed interlacing fascicles of spindle-shaped fibroblasts and an abundance of collagen without any mitotic activity (Fig 1E). The cortex of the involved ribs was thinned by pressure from the tumor, but no tumor cells were found in the ribs. The final diagnosis of posterior mediastinal fibromatosis with intraspinal and chest wall extension was confirmed. The patient recovered uneventfully and was discharged 1 week later. Neither adjuvant radiation therapy nor chemotherapy was instituted, and the patient will be followed up clinically.

Discussion

The fibromatoses are a diverse group of nonmetastasizing fibroproliferative tumors that are locally invasive and often recur after excision. Histologically, these tumors are characterized

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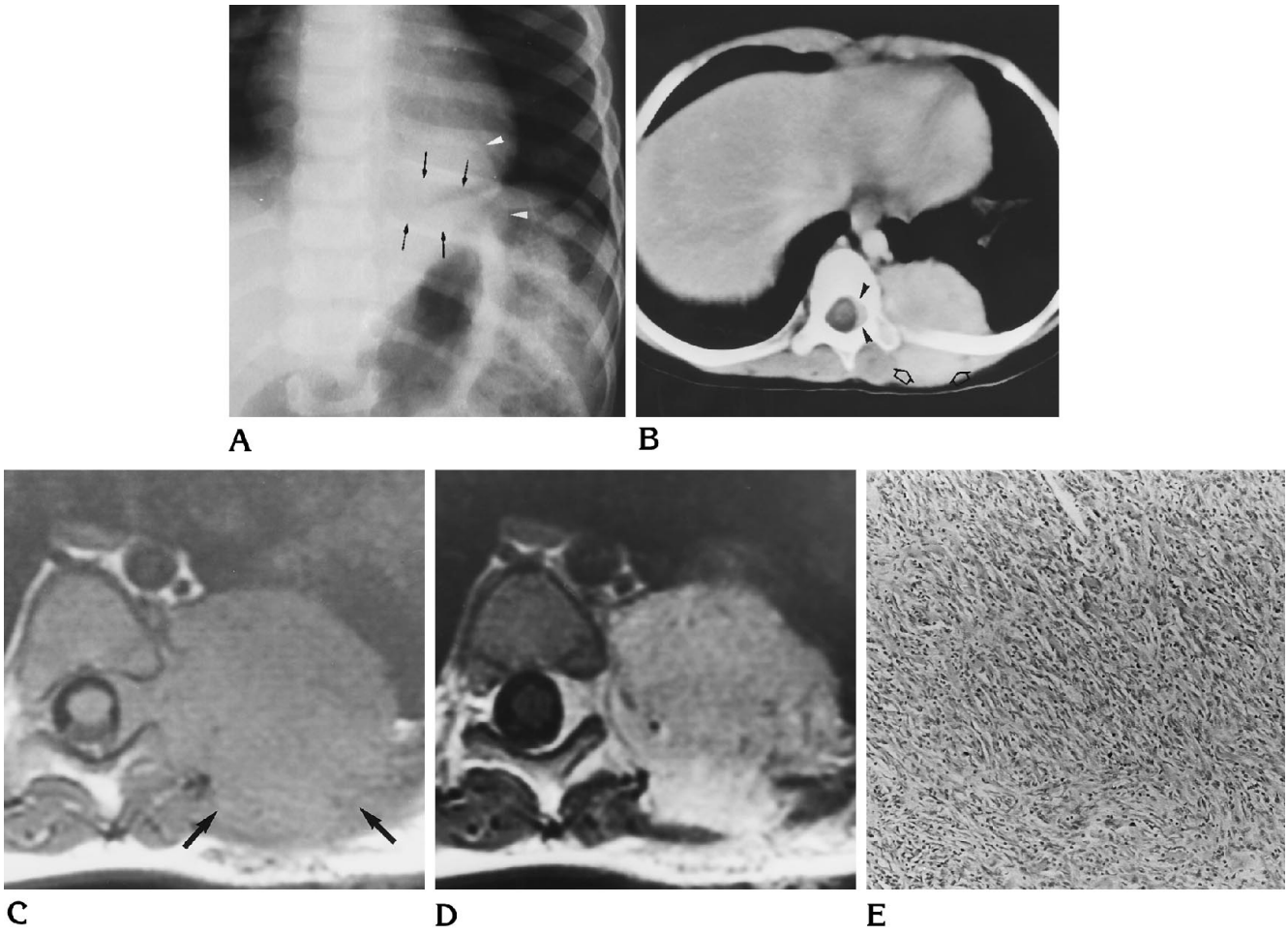

Fig 1. Posterior mediastinal fibromatosis mimicking a paravertebral neuroblastoma in a 3-year-old girl.

A, Frontal chest radiograph shows a left-sided posterior mediastinal mass (*arrows*) with erosion of the 9th and 10th ribs on the left side (*arrowheads*).

B, Contrast-enhanced CT scan shows an enhanced left paravertebral mass with direct chest wall extension (*open arrows*). The CT findings also suggest possible intraspinal invasion (*arrowheads*). No calcification is detected.

C and *D*, Unenhanced (*C*) and contrast-enhanced (*D*) axial spin-echo T1-weighted MR images (683/17) show the mass to be slightly hyperintense relative to adjacent muscle, with nearly uniform enhancement. Also shown are the chest wall invasion (*arrows*) and the transforaminal insinuation of the spine into the epidural space.

E, Photomicrograph shows a proliferation of fibroblasts in collagen stroma, which is a typical feature of fibromatosis (hematoxylin-eosin, original magnification ×330).

by fascicles of proliferating fibroblasts in a dense collagenous stroma with little or no mitotic activity (1–4). Of all neoplasms, 0.03% are abdominal fibromatoses, which occur most often in young women. The incidence of extraabdominal fibromatosis was cited in 1991 as three to four cases per 1 million people (2), and we know of one previous case (from 900 000 children) in which anterior mediastinal fibromatosis occurred after a median sternotomy for repair of a ventricular septal defect (5).

Despite the various subgroups of *adult* or *juvenile* and *superficial* or *deep* fibromatoses that have been described (1, 4), fibromatosis in children is most commonly subdivided into *congenital* and *juvenile* forms (6, 7). The congenital form is usually widespread with multiple destructive or infiltrative lesions that involve various bones and visceral organs and often lead to early death. The less aggressive juvenile form has a better prognosis and is usually confined to the musculoskeletal system (6, 7).

More than 90% of posterior mediastinal masses in children are of neurogenic origin, and the great majority of these masses are ganglion cell tumors (6–9). In our patient, the chest radiographs showed a unilateral left-sided posterior mediastinal mass. The erosion of the adja-

cent ribs indicated a slow-growing tumor. Because posterior mediastinal masses such as osteocartilaginous tumors, spinal abscess, and extramedullary hematopoiesis are usually bilateral and more destructive (6–8), they were unlikely diagnoses. CT scans depicted a solid paravertebral mass, which further excluded various cystic posterior mediastinal lesions like thoracic duct cyst, lateral meningocele, neurenteric cyst, and Bochdalek hernia (7–9). CT scans also showed that the mass extended into the chest wall and spinal canal, which was highly suggestive of a locally aggressive paraspinal neuroblastoma in this young patient (6, 7, 9). Because paraspinal neuroblastomas in children often do not contain calcification, the absence of intratumoral calcification in this case did not exclude a neuroblastoma. However, the negative results of the laboratory and bone scan studies were against the presumptive diagnosis of neuroblastoma (6–9). In retrospect, despite the rarity, we realize that the mediastinal fibromatosis was indeed the most pertinent entity that fit the clinical and imaging characteristics disclosed in this case—a noncalcified, slow-growing tumor with local infiltrative and aggressive behavior.

Most soft-tissue lesions exhibit high signal intensity equal to or greater than that of fat on T2-weighted MR images (10). Paravertebral neuroblastomas are also bright on T2-weighted images but may have a heterogeneous MR appearance depending on the presence of hemorrhage, calcification, or tumor necrosis (6, 9). Although previous case reports have suggested that hypointensity on both T1-weighted and T2-weighted images may be characteristic of aggressive fibromatosis, recent studies have supported a varying degree of signal intensity for fibromatoses in different sites, depending on the histologic composition. The greater the cellularity and the lesser the amount of collagen, the higher the T2-weighted signal intensity (2–4, 11). However, the predominant signal intensities of these lesions are quite specific: isointense to slightly hyperintense relative to skeletal muscle on T1-weighted images and typically intermediate between muscle and fat on T2-weighted images (3–4). T1-weighted images of our patient showed isointense signal intensity, but fast spin-echo T2-weighted images showed a slightly inhomogeneous left-sided paravertebral mass with signal intensity intermediate between skeletal muscle and subcutaneous fat. Furthermore, the transforaminal extradural intraspinal extension was shown to a better advantage, which greatly benefited the preoperative planning.

Wide surgical resection with adequate margin is the treatment of choice for fibromatosis. Fibromatosis is notorious for its high rate of local recurrence (12). Chemotherapy and radiation therapy have been reported to reduce the rates of local recurrence (12). However, neither treatment was appropriate for our young patient because we wanted to avoid the risks of ovarian suppression and radiation-induced spinal scoliosis. Therefore, we decided to proceed with continued clinical follow-up.

This report documents an extremely rare case of juvenile posterior mediastinal fibromatosis that mimicked a paravertebral neuroblastoma. Fibromatosis should be included in the differential diagnosis when a patient has a noncalcified, slow-growing, but locally aggressive posterior mediastinal tumor.

References

1. Allen PW. The fibromatoses: a clinicopathologic classification based on 140 cases. *Am J Surg Pathol* 1977;1:255–270
2. Quinn SF, Erickson SJ, Dee PM, et al. MR imaging in fibromatosis: results in 26 patients with pathologic correlation. *AJR Am J Roentgenol* 1991;156:539–542
3. Morrison WB, Schweitzer ME, Wapner KL, Lackman RD. Plantar fibromatosis: a benign aggressive neoplasm with a characteristic appearance on MR imaging. *Radiology* 1994;193:841–845
4. Kransdorf MJ, Jelinek JS, Moser RP, et al. Magnetic resonance appearance of fibromatosis: a report of 14 cases and review of the literature. *Skeletal Radiol* 1990;19:495–499
5. Simpson I, Campbell PE. Mediastinal masses in childhood: a review from a paediatric pathologist's point of view. *Prog Pediatr Surg* 1991;27:92–126
6. Barnes PD. Acquired abnormalities of the spine and spinal neuraxis. In: Wolpert SM, Barnes PD, eds. *MRI in Pediatric Neuroradiology*. St Louis: Mosby-Year Books, 1992:412–464
7. Swischuk LE. *Imaging of the Newborn, Infant and Young Child*. 3rd ed. Baltimore: Williams & Wilkins, 1989:189–195, 882–884, 1004–1028
8. Chapman S, Nakielny R. *Aids to Radiological Differential Diagnosis*. 2nd ed. London: Bailliere Tindall, 1990:142–143
9. Meza MP, Benson M, Slovis TL. Imaging of mediastinal masses in children. *Radiol Clin North Am* 1993;31:583–604
10. Kransdorf MJ, Jelinek JS, Moser RP, et al. Soft tissue masses: diagnosis using MR imaging. *AJR Am J Roentgenol* 1989;153:541–547
11. Lee JKT, Glazer HS. Controversy in the MR imaging appearance of fibrosis. *Radiology* 1990;177:21–22
12. Schmitt G, Mills EED, Levin V, Schmit BJ, Boeker H, Pape H. Radiotherapy of aggressive fibromatosis. *Eur J Cancer* 1992;28A:832–835