Case Report

Presacral Fibrosarcoma in Childhood: A Case Report

Warapat Virayavanich MD*, Vorachai Sirikulchayanonta MD**,
Suphaneewan Jaovisidha MD*, Suradej Hongeng MD***,
Wichian Laohacharoensombat MD****, Ratanaporn Pornkul MD*

** Department of Radiology, Faculty of Medicine Ramathibodi Hospital, Mahidol University, Bangkok, Thailand*
*** Department of Pathology, Faculty of Medicine Ramathibodi Hospital, Mahidol University, Bangkok, Thailand*
**** Department of Pediatrics, Faculty of Medicine Ramathibodi Hospital, Mahidol University, Bangkok, Thailand*
***** Department of Orthopaedics, Faculty of Medicine Ramathibodi Hospital, Mahidol University, Bangkok, Thailand*

Presacral fibrosarcoma is a rare malignant tumor in childhood. Because of its rarity in the presacral region, it is often missed from the differential diagnosis of presacral mass in childhood. The authors present a case of large presacral fibrosarcoma involving spinal canal, with an initial presentation of back pain and abnormal gaiting.

Keywords: *Childhood fibrosarcoma, Presacral mass, Sacrum*

J Med Assoc Thai 2010; 93 (2): 252-6
Full text. e-Journal: *http://www.mat.or.th/journal*

Fibrosarcoma is a tumor of the mesenchymal cell in origin that is composed of anaplastic spindle-shaped cells arranging for the most part in a fascicular (herringbone) pattern with a dense cellularity, numerous mitoses and infiltrative margins. In the past, many malignant sarcomas of other sorts have been reported as fibrosarcomas because it may be composed of what seems to be fibroblastic tissue[1]. A small biopsy specimen may fail to show the true nature of the neoplasm. Nowadays, immunohistochemical methods are often necessary to distinguish fibrosarcomas from other malignant mesenchymal tumors.

Fibrosarcoma may occur at any age but most commonly at 40-60 years[2,3], and it is uncommon in childhood. The tumor usually originates in the soft tissue of the extremities with retroperitoneal disease being relatively unusual.

Case Report

A 7-year-old girl was noted to have an abnormal gait. This abnormality was observed by her parents for 5 months before presentation. She also developed leg pain during the night. She had normal bowel habit and no urinary symptom. Past medical history was unremarkable.

Physical examination revealed a healthy girl with abnormal gait (the back deviated to the left side while walking). There was limitation of motion of the back, the hip joints, and dorsiflexion of the feet. Mild weakness of both legs was detected. No palpable mass was found in the abdominal or back region. Other systems were within normal limits. There was no significant laboratory finding.

Radiographs showed osteolytic lesions at the lumbar spine and sacral bone (Fig. 1). The abdominal ultrasonography (US) demonstrated a 5-cm well-defined hypoechoic solid mass at the presacral area without demonstrable calcification (Fig. 2). Magnetic resonance imaging (MRI) of the patient's lumbosacral spine showed a large mass filling the presacral region, obstructing the left ureter and causing hydronephrosis of the ipsilateral kidney (Fig 3A). On the sagittal MR images, the mass had destroyed the S2, S1, and L5 vertebral bodies and extended into the central spinal canal up to lower L4 level, resulting in severe thecal sac and multiple nerve roots compression from L4 and downward (Fig. 3B, C). The lesion showed isosignal intensity on T1-weighted images and slightly inhomogenously high signal intensity on T2-weighted images. Gadolinium-enhanced MR study showed a

Correspondence to: Pornkul R, Department of Radiology, Faculty of Medicine, Ramathibodi Hospital, Mahidol University, Rama 6 street, Bangkok 10400, Thailand. Phone: 0-2201-1212, Fax: 0-2201-1297

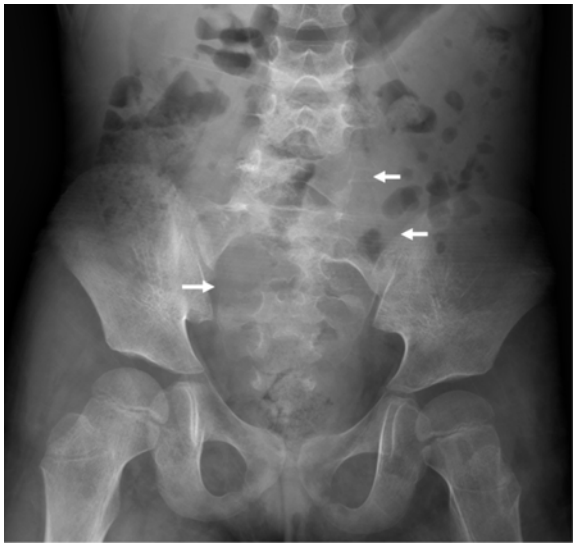

Fig. 1 Pelvic radiograph shows osseous destruction at both sides of sacrum and left side of L5 (arrows)

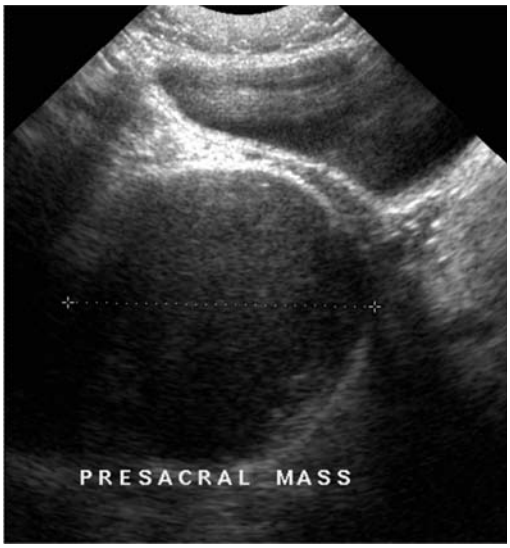

Fig. 2 Transverse ultrasound image reveals a solid presacral mass (measured) causing pressure effect to the urinary bladder

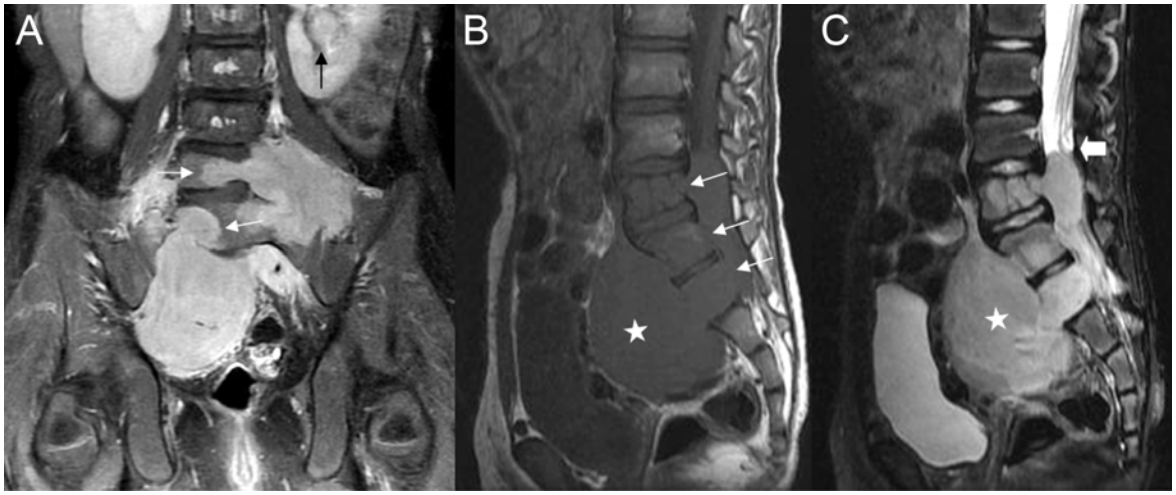

Fig. 3 MR imaging of the presacral mass. (A) coronal T1 fat-suppressed after Gadolinium injection, (B) sagittal T1 and, (C) sagittal T2 images reveal a large mass occupying presacral area (star), destroying the sacrum and L5 vertebra (thin white arrows in [A] and [B]), and filling the spinal canal up to lowerL4 level (thick white arrow in [C]). This mass causes hydronephrosis in the left kidney (thin black arrow in [A])

rather homogeneous enhancement in the mass. The bone scan revealed a photopenic area at the left-sided L5 vertebra and right-sided sacrum, corresponding to the mass (Fig. 4).

Core needle biopsy at right-sided S1 failed. Then, a mini-opened biopsy was done via a 2-cm skin incision and the tumor tissue was harvested from the posterior aspect of the right pedicle of the S1 vertebra by pituitary forceps. The histological study showed cellular and slightly pleomorphic spindle cells having enlarged vesicular fusiform nuclei, basophilic nucleoli, and slender eosinophilic cytoplasms running in interlacing bundles and occasional storiform-pattern (Fig. 5, 6). Some foci showed presence of islands of bone trabeculae surrounded by tumor cells. Few mitotic figures per high-power fields were recognized

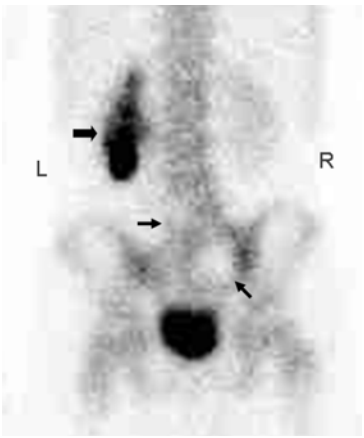

Fig. 4 Posterior image of planar bone scan shows photopenic area surrounding with increased uptake at right-sided sacrum, and photopenic area at left side of L5 (thin arrows). Evidence of isotope accumulation observed in the left kidney is due to hydronephrosis (thick arrow)

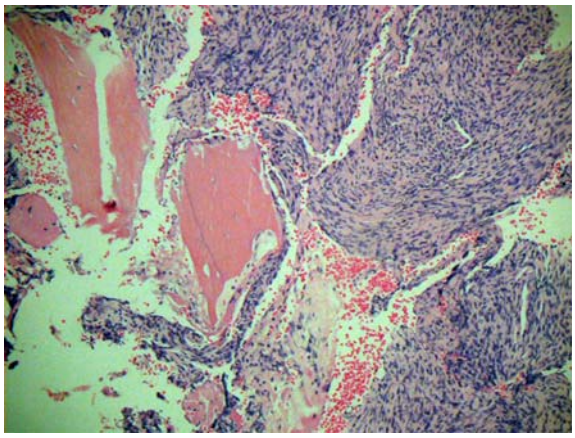

Fig. 5 (H&E; x100) Photomicrograph reveals tumor entrapping islands of bone-trabeculae and made up of cellular spindles running in interlacing bundles

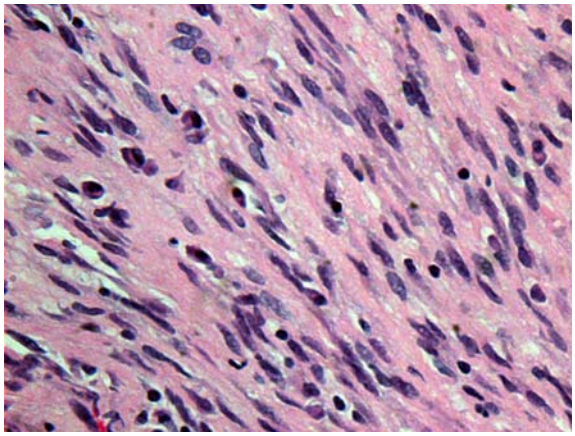

Fig. 6 (H&E; x400) Photomicrograph shows that spindle cells possess enlarged, slightly pleomorphic vesicular nuclei with small basophilic nuclei and slender eosinophilic cytoplasms

in the active fields. No appreciable osteoid matrix was observed. The differential diagnosis included fibroblastic type of osteosarcoma, leiomyosarcoma, and fibrosarcoma. However, the immuno-stains revealed that the tumor cells had negative reactivity to desmin, actin (SM), and osteocalcin. The diagnosis of fibrosarcoma, low-grade was therefore entertained.

There was no evidence of bony or pulmonary metastasis at the time of diagnosis.

The patient was treated with systemic combination chemotherapy, debulging tumor, and palliative radiotherapy. Repeated MRI showed slight reduction of the size of the mass.

Discussion

The presacral retroperitoneal space is a potential space between perirectal fascia and the fibrous tissue covering the anterior sacrum. The lateral margins are defined by the ureters and the iliac vessels. It is an area where the neuroectoderm, notochord, hindgut and protoderm undergo remodeling and regression in embryological life. It can thus be the site of a heterogeneous group of benign and malignant tumors.

Tumors in the presacral area are rare, and the true incidence is difficult to assess. Jao et al[4] reported on 120 patients with presacral tumors treated at the Mayo Clinic during a period of 19 years. Those tumors represented 1 in every 40,000 general hospital admissions. Certain studies have been published involving a large number of presacral malignant tumors; however, presacral fibrosarcoma were not identified in any of these studies[5,6].

Childhood fibrosarcoma is histologically similar to classic adult fibrosarcoma but it is considered to hold a favorable prognosis when compared to the adult form[7]. The tumors form 3% of all cases of malignant tumors in infancy and childhood[8]. Childhood fibrosarcoma presenting as a retroperitoneal mass is rare and only a few cases have been reported so far[9-12]. Soule et al[13] reviewed

110 cases of childhood fibrosarcoma and found that about 62% of cases were diagnosed in the first quinquennium (birth to 5 years), 12% in the second, and 26% in the third, and most of the tumors were situated in the extremities. Only eight of the 110 patients had fibrosarcoma located in the trunk. However, they did not specify how many of these cases were located in the retroperitoneum.

Of the 134 cases of fibrosarcoma in infants and children (under 15 years of age) in the study of Kransdorf[14], only six (4%) were retroperitoneal. This malignancy chiefly affected the lower and upper extremities.

The symptoms of a mass in the presacral space depend on both the location and the size of the mass [3, 6]. Because the presacral retroperitoneal space is a potential space, it is the possible reason why these tumors can develop to such a large size before symptoms occur clinically. Most patients have non-specific symptoms; included are changes in bowel habits, low back and sacral pain, abdominal pain, palpable pelvic or abdominal mass, and urinary symptom[6]. Because the clinical manifestations of presacral masses are often non-specific, imaging therefore plays an important role in the detection and differentiation of these masses.

The imaging features of fibrosarcomas are non-specific. Radiological examinations such as conventional radiography and gastrointestinal and urinary contrast material-enhanced studies are usually unsatisfied. Instead, the US, MRI, and computed tomography are considerably used in the evaluation of presacral masses by performing alone or in combination[6,15-17]. Multiplanar MRI is extremely useful in the evaluation of sacral and presacral lesions, and having great value in management planning because it accurately demonstrates both soft tissue and intrasacral extent to determine possible resectability. In addition, MRI can help characterize the lesion as solid, cystic, fatty, and hemorrhagic.

A characteristic feature of childhood fibrosarcoma is a high local recurrent rate of 33-43%, but the development of metastasis is rare[13,18]. Overall survival is about 90% at 5 years[12,18]. Recurrence of childhood fibrosarcoma has been reported as late as 31 years following the initial operation[11].

The mainstay of treatment consists of surgical resection by wide excision; however, a majority of patients has unresectable tumors at diagnosis. More recently, combination chemotherapy has given good results, with the effect that various imaging modalities have become important in assessing both the initial extent of disease and the response to treatment[11,19,20].

Conclusion

The authors presented a rare case of fibrosarcoma that originated in the presacral space of a child. Because of its rarity, it is usually missed from the differential diagnosis of presacral mass in childhood. The diagnosis established by biopsy of a specimen with exclusion of other tumors by immuno-histochemical methods. Because presacral space is a potential space, the mass may grow up to a large size before the patient seeks medical treatment. Due to the clinical symptoms being non-specific, imaging particularly the MRI plays an important role in multiple purposes.

References

1. Stout AP. Fibrosarcoma the malignant tumor of fibroblasts. Cancer 1948; 1: 30-63.
2. Pritchard DJ, Soule EH, Taylor WF, Ivins JC. Fibrosarcoma-a clinicopathologic and statistical study of 199 tumors of the soft tissues of the extremities and trunk. Cancer 1974; 33: 888-97.
3. van der Werf-Messing B, van Unnik J. Fibrosarcoma of the soft tissue: a clinicopathologic study. Cancer 1965; 18: 1113-23.
4. Jao SW, Beart RW Jr, Spencer RJ, Reiman HM, Ilstrup DM. Retrorectal tumors. Mayo Clinic experience, 1960-1979. Dis Colon Rectum 1985; 28: 644-52.
5. Cody HS 3rd, Marcove RC, Quan SH. Malignant retrorectal tumors: 28 years' experience at Memorial Sloan-Kettering Cancer Center. Dis Colon Rectum 1981; 24: 501-6.
6. Soye I, Levine E, Batnitzky S, Price HI. Computed tomography of sacral and presacral lesions. Neuroradiology 1982; 24: 71-6.
7. Stout AP. Fibrosarcoma in infants and children. Cancer 1962; 15: 1028-40.
8. AndersenNDERSEN DH. Tumors of infancy and childhood. I. A survey of those seen in the pathology laboratory of the Babies Hospital during the years 1935-1950. Cancer 1951; 4: 890-906.
9. Hamm CM, Pyesmany A, Resch L. Case report: congenital retroperitoneal fibrosarcoma. Med Pediatr Oncol 1997; 28: 65-8.
10. Trobs R, Meier T, Bennek J, Heinrich S, Willnow U. Fibrosarcoma in infants and children: a retrospective analysis - overdiagnosis in earlier

years. Pediatr Surg Int 1999; 15: 123-8.
11. Wong CH, Dietrich RB, Pais MJ, Zeltzer PM, Carpenter PM. Pediatric case of the day. Congenital, undifferentiated retroperitoneal fibrosarcoma. Radiographics 1994; 14: 1428-30.
12. Chung EB, Enzinger FM. Infantile fibrosarcoma. Cancer 1976; 38: 729-39.
13. Soule EH, Pritchard DJ. Fibrosarcoma in infants and children: a review of 110 cases. Cancer 1977; 40: 1711-21.
14. Kransdorf MJ. Malignant soft-tissue tumors in a large referral population: distribution of diagnoses by age, sex, and location. AJR Am J Roentgenol 1995; 164: 129-34.
15. Kocaoglu M, Frush DP. Pediatric presacral masses. Radiographics 2006; 26: 833-57.
16. Diel J, Ortiz O, Losada RA, Price DB, Hayt MW, Katz DS. The sacrum: pathologic spectrum, multimodality imaging, and subspecialty approach. Radiographics 2001; 21: 83-104.
17. Eich GF, Hoeffel JC, Tschappeler H, Gassner I, Willi UV. Fibrous tumours in children: imaging features of a heterogeneous group of disorders. Pediatr Radiol 1998; 28: 500-9.
18. Cofer BR, Vescio PJ, Wiener ES. Infantile fibrosarcoma: complete excision is the appropriate treatment. Ann Surg Oncol 1996; 3: 159-61.
19. Loh ML, Ahn P, Perez-Atayde AR, Gebhardt MC, Shamberger RC, Grier HE. Treatment of infantile fibrosarcoma with chemotherapy and surgery: results from the Dana-Farber Cancer Institute and Children's Hospital, Boston. J Pediatr Hematol Oncol 2002; 24: 722-6.
20. Grier HE, Perez-Atayde AR, Weinstein HJ. Chemotherapy for inoperable infantile fibrosarcoma. Cancer 1985; 56: 1507-10.

เนื้องอก fibrosarcoma ที่ตำแหน่งหน้าต่อกระดูกใต้กระเบนเหน็บในผู้ป่วยเด็ก: รายงานผู้ป่วย 1 ราย

วราพัฒน์ วิระยะวานิช, วรชัย ศิริกุลชยานนท์, สุภนีวรรณ เชาว์วิศิษฐ, สุรเดช หงส์อิง, วิเชียร เลาหเจริญสมบัติ, รัตนพร พรกุล

เนื้องอกชนิด fibrosarcoma เป็นเนื้องอกชนิดร้ายที่พบได้น้อยมากที่ตำแหน่งหน้าต่อกระดูกใต้กระเบนเหน็บในผู้ป่วยเด็ก และเนื่องจากการที่เนื้องอกชนิดนี้พบได้น้อยในตำแหน่งนี้ ทำให้การวินิจฉัยโรคนี้ในผู้ป่วยเด็กที่มาด้วยก้อนที่อยู่หน้าต่อกระดูกใต้กระเบนเหน็บมักถูกมองข้าม ผู้นิพนธ์ได้รายงานผู้ป่วย 1 ราย ที่ตรวจพบก้อนขนาดใหญ่อยู่หน้าต่อกระดูกใต้กระเบนเหน็บ และโตเข้าไปในกระดูกใต้กระเบนเหน็บและโพรงกระดูกสันหลัง ทำให้ผู้ป่วยมาพบแพทย์ด้วยอาการปวดหลังและท่าเดินผิดปกติ