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SYNOVIAL SARCOMA IN CHILDHOOD: CLINICAL AND RADIOLOGICAL FINDINGS

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Objective: To study the clinical characteristics and radiological features of synovial sarcoma in childhood and its relation to the diagnosis and treatment. Methods: The clinical radiological features of 15 children with synovial sarcoma proved surgically and pathologically were analyzed. Results: In children, the tumor boundaries are poorly defined due to paucity of fat, and metastasis usually occurs early. Eight patients in this series had bone involvement, including: direct erosion by tumor causing cortical destruction, indirect pressure defect with sharp margin and reactive bone sclerosis and bone destruction of the primary intraosseous synovial sarcoma. Conclusion: The tumor is often misdiagnosed, the final confirmed diagnosis must be made by histological examination with imaging findings. It is emphasized that the patients should be treated with radiotherapy and chemotherapy preoperatively and postoperatively.

Key words: Synovial sarcoma, Radiograph, Diagnosis, Surgery, Radiotherapy, Chemotherapy

Synovial sarcoma is an uncommon malignant mesenchymal tumor, accounting for about 2 percent of primary malignant soft-tissue tumors.[1] This tumor is mostly seen in adults, only 13 percent occurred in childhood.[2] Misdiagnosis is common because of the absence of typical clinical and radiological features and postoperative recurrence and early metastases are frequently found.[2-10] Recently, the major focus of the tumor is on the pediatric patients,[11-13] but no report focused on it can be found in Chinese literature. We here presented 15 cases of histologically proved synovial sarcoma in childhood and discussed related issues.

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MATERIALS AND METHODS

The cases came from Shandong, Jiangsu, Beijing, Hebei, Henan, Sichuan, Hunan, etc. 10 of them were one part of a series of 115 patients with synovial sarcoma.[2] All 15 cases had plain films of involved area. Six of the cases with lower extremity involvement underwent angiography. CT scan was performed in 2 cases.

RESULTS

Clinical Presentations

There were 9 males and 6 females. The ages ranged from 4 to 14 years with an average of 10.8 years. The mean course was 7.3 months with a range of 2 months to 2 years. The durations of symptoms worsening were from 2 days (trauma) to 35 days.

The main presentation was a local mass with or without pain. The size of masses ranged from 4×3 to 14×10cm, in 12 cases the diameter of tumor was over 6 cm. Other complaints included limp, limitation of joint movement, or chest pain if chestwall was involved, as well as symptoms of metastatic disease.

Four of the 15 cases had pulmonary metastases before biopsy and were alive for 24 days to 3 months

after diagnosis, and one of them had extensive bone metastases. The rest 11 patients (including 3 cases with local lymphatic metastases) underwent local excision (8 cases) or amputation (3 cases), and seven had local recurrence postoperation 2 to 11 months after operation, including four with local lymphatic metastases. Only 2 patients were survived 2 years after operation.

Pathologic Findings

Grossly the tumor was fish-fleshy and fragile, some were tough. Necrotic area was frequently found in the center of a big tumor. Microscopic examination revealed characteristic diphasic differentiation with both epithelioid cells and spindle fibrocytes, and glandular ducts like crevices were also shown (Figure 1).

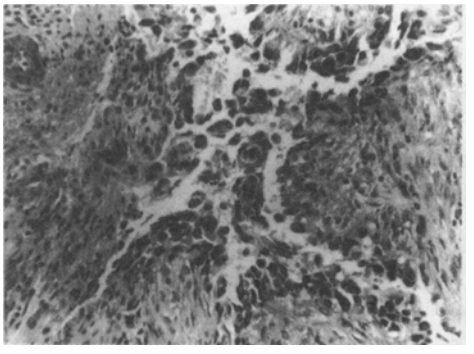

Fig. 1. The pathological findings of synovial sarcoma. Diphasic differentiation of tumor cells and glandular duct like crevices were shown.

Radiologic Manifestations

Plain Radiograph:

(1) Soft Tissue Mass: It was found in all 15 cases and presented as watery density. When the tumor located in the area with rich soft tissue, its margin was poorly defined (Figure 2); while in the area with thin soft tissue, the tumor might has a relative clear margin that was lobular (Figure 3) or smooth, but intermuscular spaces and subdermal fat were indistinct.

(2) Osseous Changes: Eight cases had osseous changes, including 2 cases with direct invasion of underlying bones presenting cortical destruction, 3 cases with pressure erosion (Figure 4) and reactive sclerosis (Figure 5), 2 cases with primary intra-osseous destruction (Figure 6), and 1 cases with osteoporosis.

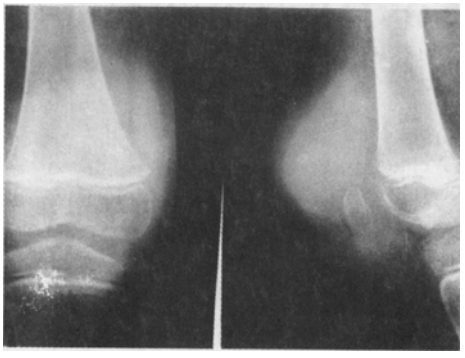

Fig. 2. The tumor located in proximal thigh. Its margin was poorly defined.

(3) Joint space and articular surfaces: In 10 cases, synovial sarcoma was located in the vicinity of joint, including 2 cases with a great soft-tissue lump, whose articular surfaces were undamaged. Osteoporosis might be seen in the long-course cases. Intact joint spaces were usually normal (Figure 3).

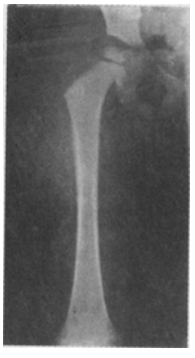

Fig. 3. Synovial sarcoma in a knee. The lobular tumor mass had a relatively clear margin. The joint space and articular surfaces were normal.

(4) Calcification of tumor: Only 2 cases showed calcification in the tumor mass.

Angiography:

In six patients who had under went angiography, tumors were all hypervascular with irregular and large blood vessels in their periphery, and the adjacent blood vessels were pushed. Early display of veins was found in 2 cases and "tumor staining" in one case.

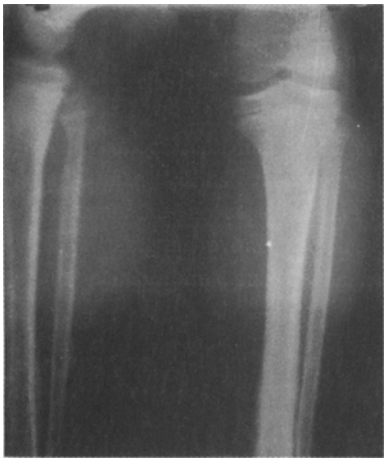

Fig. 4. The tumor involved a leg. The axially narrowed tibia was shown. A depression in proximal fibula was also noted.

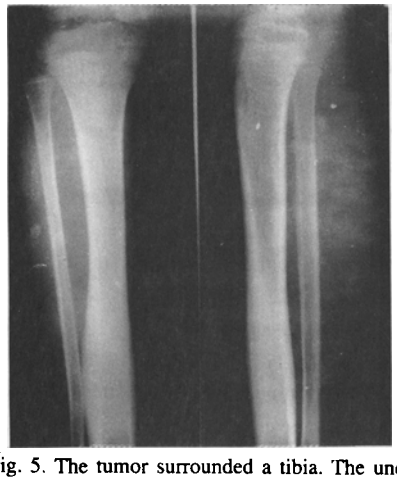

Fig. 5. The tumor surrounded a tibia. The underlying proximal-middle tibia narrowed axially with thinned cortex, while the distal tibia widened with thickened and sclerotic cortex.

CT: In 2 cases (including one with a tumor exceeding 10 cm in diameter) CT failed to reveal the exact margins of tumors, while it demonstrated necrotic area and faint calcification in the tumors clearly.

DISCUSSION

Synovial sarcoma is a malignant tumor that is more harmful than Wilms' tumor for childhood. Because the tumor has no capsule[13] and spreads by direct extension along the myofascial planes, it usually does not cause pain or form a mass. When a notable

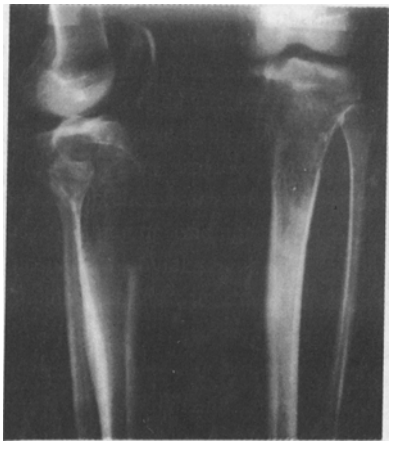

Fig. 6. Intraosseous synovial sarcoma. The osteolytic destruction of proximal tibia with pathological fracture was shown. The slight expansive lesion contained dense spots. No periosteal reaction was found.

mass emerges associated with pain or other symptoms, the tumor is often larger than 6 cm in diameter and distal metastases are common. Four cases of the series had pulmonary metastases before biopsy and all died within 3 months after diagnosis. The natural survival duration ranged from 6 to 14 months with an average of 10.5 months. In 11 cases treated with operation, two years survival rate was only 18%, which was much lower than the five years survival rate of 38%—45% in adult patients.[6,9]

1. Diagnosis

In the early stage, the tumor is often misdiagnosed as benign disease or normal due to the absence of typical symptom or sign, even if imaging examinations have been done. Soft tissue mass has been thought to be a sole sign of synovial sarcoma in childhood.[12] In this series, most large tumors were associated with osseous changes such as cortical destruction, pressure erosion and reactive sclerosis. Extensive calcification had been suggested to indicate a more favorable prognosis,[5] while only 2 cases showed calcification in the tumor mass. Although CT can easily define the extent and size of the masses, it is difficult to assess the exact margin of the tumors in children because of the relative paucity of fat.[12] Angiography may reveal the blood supply of the tumor but can not provide diagnostic features. Therefore, comprehensive analysis of clinical presentations and imaging findings is most important in the diagnosis of synovial sarcoma in childhood. The imaging manifestations with diagnostic value include

a soft-tissue mass in the vicinity of joints, especially of large joints in lower extremities, containing calcification, and the pressure erosion and reactive sclerosis of under lying bone. Clinically, the chronic and progressive features are helpful in considering the diagnosis of the tumor. The final confirmed diagnosis must be made by histological examination.

2. Treatment

When considering the treatment of synovial sarcoma in childhood with surgical excision, it must be kept in mind that the tumor spreads by direct extension along the myofascial planes and a wider erasion should be the first choice. In Israels' series, a six years old patient with adjacent invasive synovial sarcoma was treated with a wider excision and was alive 27 years after diagnosis; two patients with high-grade malignant tumor had amputation and were alive 4 and 8 years respectively.[12]

In the past, the major focus of treatment of synovial sarcoma has been surgical.[11] Recently, the "limb-salvage" procedures has been emphasized. This approach includes irradiation to the involved area before and after wide local excision of the tumor and adjutant chemotherapy after operation.[7,8] A 12-year-old patient with synovial sarcoma of high-grade malignancy received 5,000 rad (50Gy) radiotherapy in 25 fractions and chemotherapy (vincristine, cyclophosphamide, adriamycin, and high-dose methotrexate) after surgical excision of the tumor and adjacent muscles when microscopic tumor was found at the deep resection margin of the muscles. The patient had no evidence of tumor recurrence 15 months later.[12] For the cases with tumor recurrence after local excision, Waag and co-workers applied a wider excision preceded by chemotherapy and radiotherapy (50 Gy), and satisfactory results were achieved.[13] More recently, electrochemotherapy, a new non-invasive and non-toxic method, has been advocated, it may be a more ideal method for synovial sarcoma.

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