Fibrosarcoma in Children

A Rare Tumour with Long-Term Survival even with Advanced Disease

A Report of 3 Cases

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Fibrosarcoma is a rare tumour in children. The potential of malignancy has been questioned. We present three cases of fibrosarcoma in children . The follow-up periods range from 10 to 37 years. The first patient had pulmonary metastases at the time of diagnosis in 1958. The primary tumour in fossa ischio-rectalis was resected in 1960. Lung metastases were resected in 1960 and 1989. Radiotherapy was given in 1992. He is still alive with metastases 37 years after the first manifestation of disease. The second patient had a primary tumour and several local recurrences in the mandible. He is alive without evidence of disease 4 years after resection of pulmonary metastases and 21 years after resection of the primary tumour. The third patient has no signs of recurrence or metastasic spread 10 years after a wide excision of subcutanous tumours of the left upper arm. The cases demonstrate a special tumour-entity of low-grade malignancy, which show a good prognosis and a wide spectrum of biological behaviour.

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Fibrosarcoma in children usually behave in a more favourable way than their adult counterpart (1). The most common localizations are the distal parts of the extremities. The trunk, head and neck regions are less commonly involved (2). Some authors (3) consider fibrosarcoma a borderline tumour because of its infrequently manifested potential for metastasis; in Coffin & Dehners (3) material none out of 13 cases behaved in a malignant fashion. Some (4) even maintain that designating these tumours as sarcomas is a misnomer. Others (5), however, report a rather poor prognosis in infants less than 1 year of age. Enzinger & Weiss (2) found a 5-year survival rate of 84% in their series of infantile fibrosarcomas.

We report 3 cases of fibrosarcoma in children, with long-term survival.

Case 1. A 13-year-old boy presented in 1958 with an egg-sized tumour in fossa ischio-rectalis and radiographs revealed tumours in his left lung. The soft tissue tumour and the tumours in his lung were growing slowly, and they were all removed surgically in 1960. Pulmonary metastasis recurred and became bilateral in 1971. The disease progressed slowly. Because of respiratory symptoms some of the tumours in the left lung were resected in 1989. The histological picture was almost the same as in 1960 (Fig. 1).

In 1992, he was given radiotherapy as symptomatic treatment for central pulmonary metastases. In 1995, 37 years after the first manifestation of disease, the patient was still alive and in relatively well being with the help of analgesics. He presented bilateral pulmonary and mediastinal metastases.

Case 2 . A 2.5-year-old boy was referred to the Department of Oral and Maxillo-Facial Surgery, Haukeland University Hospital in 1974. Following a trauma to his face, his mother had observed a rapidly growing mass in the region of the boy's right mandible.

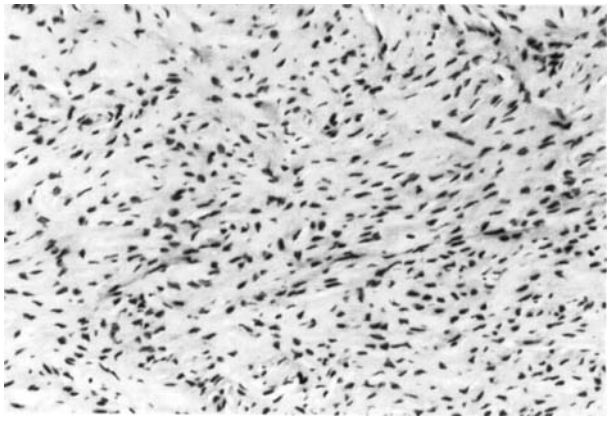

Fig. 1. Biopsy of pulmonary metastasis 30 years after the primary tumour was discovered. The cells are spindle-shaped and growing in ill-defined fascicles. There is only slight nuclear variation and no mitoses. HES × 56.

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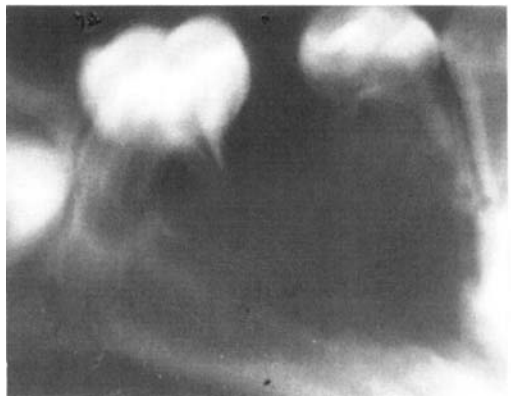

Fig. 2. Two and a half-yr old boy with mandibular infantile fibrosarcoma. Preoperative radiograph showing displacement of tooth bud.

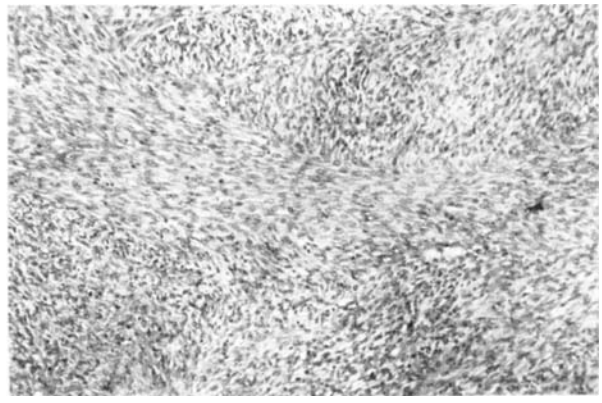

Fig. 3. Infantile fibrosarcoma. Spindle-shaped cells with the classic fascicular arrangement. Some pleomorphism, no mitoses. HES × 56.

The x-ray examination revealed a radiolucent ill-defined lesion in this region, with extensive resorption of teeth 85 and 84 and displacement of tooth bud 45 (Fig. 2). The tumour was enucleated in 1974 and the histologic picture of the tumour is shown in Fig. 3. Later the same year a wide excision and reconstruction of the mandible using rib graft was performed. During the years 1975, 1979, 1980 and 1981 he presented local recurrences which were treated with local excisions. There was no history of relapse until 1991 when a local recurrence and also a solitary pulmonary metastasis occurred. Both tumours were surgically removed and the histologic picture showed the same morphology as in 1974. In 1994 he experienced yet another recurrence which was treated with marginal excision and radiotherapy. The patient is still alive, with no evidence of disease, 21 years after the primary tumour was discovered.

Case 3. A 3-month-old boy presented in 1985 with 2 subcutanous tumours located superior to the biceps muscle of his left arm. Both tumours were surgically removed and the histologic picture is shown in Fig. 4. Two months later, a wide surgical excision was performed. There has been no local recurrences or signs of metastasis in the 10-year follow-up period.

Histopathology. Tissue from all tumours were prepared with routine histological procedures. The samples were embedded in paraffine, sectioned at 5 μm and stained with hematoxylin and eosin. Immunoperoxidase reactions for vimentin, desmin and S-100 has been performed in all 3 cases.

All the tumour specimens presented a histologic picture with cellular immature-appearing fibroblasts growing in a fascicular arrangement (Figs. 1, 3, 4). The tumour cells were spindle-shaped and the mitotic activity was low. The cells stained positive to vimentin, while desmin and S-100 gave negative results. In all three cases the histopathologic diagnosis was fibrosarcoma of low grade malignancy.

Discussion. In one of the three cases the primary tumour had the typical location of fibrosarcomas, namely in the distal part of an extremity. The other cases had less common localizations; in the buttock region and in the mandible.

Histologically, these tumours usually are composed of solidly packed, spindle-shaped cells separated by variable amounts of interstitial collagen (6). Different growth patterns can be seen, the fascicular or the herringbone-like being the most classical one. Infiltration of inflammatory cells, myxoid- and haemangiopericytoma-like areas are also described (7). Multinucleated giant cells are rare, whereas mitotic figures are common, although their number varies from case to case and even in different portions of the same tumour (2).

The histopathologic diagnosis were fibrosarcomas in all 3 cases reported, 2 of them classified as infantile fibrosarcomas.

The follow-up periods differ from 10 to 37 years. The two patients with the longest follow-up periods have both advanced disease with pulmonary metastases while the third patient has had no signs of metastatic disease after 10 years. The survival times with pulmonary spread are 4 and 37 years respectively, and the patients are still alive which indicates a low malignant biological behaviour.

The treatment and the use of chemotherapy for this tumour are controversial. Late presenting side-effects, including development of a second malignant tumour, must be considered before treating children with chemotherapy. Some centres (8, 9), have used chemotherapy preoperatively in selected cases. However, the treatment of choice is usually a wide excision. Radiotherapy and chemotherapy should be reserved for patients with inoperable tumours and for children in whom curative surgery would be mutilating (8, 9). None of the patients referred to in this article received chemotherapy. One patient experienced long term palliation following radiotherapy of lung metastases. However, it is too early to say whether radiotherapy has influenced the tendency to local recurrences in this patient who was given radiotherapy postoperatively to the premandible and submandibular regions after excision of a local recurrence. In one case, the pulmonary metastases were considered a malignant fibrous histiocytoma of low-grade malignancy, 30 years after the primary tumour was discovered. This finding has also been reported by others (5).

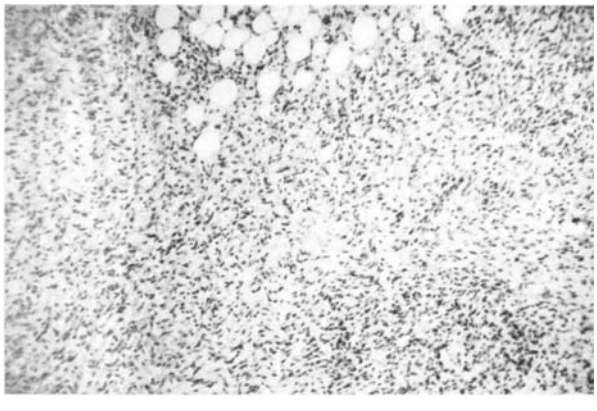

Fig. 4. Infantile fibrosarcoma with myxoid areas of spindle-shaped cells infiltrating subcutaneous tissue. HES × 56.

Some authors (7) recommend fluorescence in-situ hybridization or cytogenetics to determine chromosome 8, 11, 17 and 20 copy numbers in these tumours. This may be helpful, since the histologic diagnosis can be very difficult, even for trained pathologists. Unfortunately, we are not yet in a position to perform such studies at our laboratory. Because of limited surgical experience regarding these tumors, we recommend that these patients are referred to large hospitals.

Conclusion. Fibrosarcoma in children is a rare tumour of low grade malignancy. The histopathologic diagnosis might be difficult to establish, even for trained pathologists. The tumour has a wide spectrum of biological behaviour. Long-term survival can be seen, also in patients who present with local recurrences and metastases.

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