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An Infantile Low Grade Fibromyxosarcoma Of The Neck: A Rare Case

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ABSTRACT

Background: *Fibrosarcoma is a mesenchymal tumor of adult and pediatric age groups representing 10 to 12% of all soft-tissue sarcomas. Infantile fibrosarcoma is an uncommon tumor of childhood. It represents less than 1% of all childhood cancers but is the most common soft-tissue sarcoma in those under 1 year of age. The tumor may be observed at birth, in infancy or in childhood, and has been called congenital or infantile fibrosarcoma. Fibrosarcoma is one of the types of non-rhabdomyosarcoma soft tissue sarcoma (NRTS) that accounts for 3% of all childhood malignancies. Only 5% of all primary malignant tumors in children arise in the head and neck. The purpose of this paper is to report a rare case of infantile low grade fibromyxosarcoma and the choice of diagnostic radiology modalities.*

Case: *A 2-month-old boy with the rare case of an infantile low-grade fibromyxoid sarcoma (LGFMS). The tumor arose in the paravertebral region of his left posterolateral neck. There is no infiltration to the intracranial or vertebral canal. A review of the literature indicates that this is one of the youngest patients diagnosed with a tumor of this type, and had never been reported as LGFMS.*

Conclusion: *A rare case of infantile low grade fibromyxosarcoma had been reported. Based on the history, physical examination, laboratory examination, and radiologic imaging, the diagnosis of infantile low grade fibromyxosarcoma was established. Magnetic Resonance Imaging (MRI) is the best modality overall for examining soft-tissue masses and is useful in providing information about the local extent, lesion size, and involvement of the neurovascular structures.*

Keywords: *Fibromyxoid, sarcoma, childhood, radiology*

INTRODUCTION

Infantile fibrosarcoma is an uncommon tumor of childhood. Infantile fibrosarcoma represents less than 1% of all childhood cancers, but it is the most common soft-tissue sarcoma in those under 1 year of age. Fibrosarcoma is one of the types of non-rhabdomyosarcoma soft tissue sarcoma (NRTS) that accounts for 3% of all childhood malignancies. Only 5% of all primary malignant tumors in children arise in the head and neck.[1,2]

Fibrosarcoma is a tumor of mesenchymal cell origin that is composed of malignant fibroblasts in a collagen background. It can occur as a soft-tissue mass or as a primary or secondary bone tumor. Low-grade fibromyxoid sarcoma (LGFMS) is a rare soft tissue sarcoma that typically affects young adults, equally in men and women (median age at presentation 34 years). It mostly locates in the deep soft tissue, presenting as a slow-growing, nontender, firm tumor. However, LGFMSs also occur in superficial regions and in children more often than previously recognized.[3]

Some subtypes of pediatric NRSTS (e.g., infantile fibrosarcoma) have a relatively benign clinical course after treatment with surgery alone. NRSTS, like rhabdomyosarcoma, can arise in any part of the body. Two series, comprising a total of 322 pediatric patients, revealed that the most common sites were the extremities, trunk, and abdomen and pelvis. In these series, synovial sarcoma, neurofibrosarcoma, malignant fibrous histiocytoma, and fibrosarcoma were the most frequent histologies.[4]

Nonrhabdomyosarcomatous soft tissue sarcomas (NRSTS) occurring in children are a diverse group of neoplasms whose rarity has hindered elucidation of the optimal treatment and prognosis. These tumors are more frequently encountered in adults, and data about their impact on children are extrapolated from clinical studies that involved an older patient population.[5]

The etiology of cancer in children still is poorly understood, for the most part, epidemiology studies have recognized that the likely mechanism is multifactorial, possibly resulting from potential interaction between genetic susceptibility traits and environmental exposures.[6]

Because the tumor is so rare, clinical experience is hacking, and there is no established protocol for comparing the effectiveness of operative treatment, chemotherapy, and radiation. Treatment must be selected on an individual basis. Although complete resection is rarely feasible at diagnosis, conservative surgery remains the mainstay treatment for infantile fibrosarcoma. An alkylating agent-free and anthracycline-free regimen is usually effective and should be chosen as first-line chemotherapy for inoperable tumors.[1,7]

Overall prognosis is good, but progression or relapse, mainly local, remains possible. Infantile fibrosarcoma , unlike the adults, has a good prognosis when treated with a combination of resection and chemotherapy.[7]

The purpose of this paper is to report a rare case of infantile low grade fibromyxosarcoma and the choice of diagnostic radiology modalities.

CASE REPORT

AR, 2-month-old baby boy was referred to the Surgery Department with a mass in the side of his left neck. The mass was noticed since birth. There was a slight enlargement of the mass. He had no history of fever, decreased of body weight, bleeding and there was no respiratory distress nor difficulty of feeding.

History of birth and pregnancy. He was born by Caesarean section, was 50 cm in length, 4000 grams in weight (with the mass). He was the first child of the family. His mother's age was 22 years old, with no history of illness during pregnancy. She had control routine check-ups from the local midwife every month. At 8 months of pregnancy she underwent an ultrasound examination and was told by the doctor that there was a mass on the fetus' neck.

Family illness history: There was no history of malignancies in either his mother or his father.

Physical examination. The vital signs were within normal limit. There was no respiratory distress, enlargement of the lymph nodes, liver or spleen. The mass on the left neck was firm, not tender and immobile. The overlying skin was tight and with normal color. The mass size was about 17 cm in diameter. There were no palpable masses in other parts of the body.

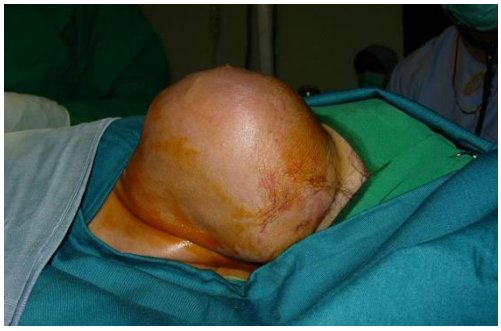

Figure 1. The mass on the left neck

The anthopometric measurements revealed a good nutritional state and he weighed 5800 grams and his length was 52 cm. The laboratory findings Hb 11,1 WBC 10.900, platelet count 393.000. SGOT/SGPT: 34/50 Ur/Cr: 9,7/0,13. Albumin: 3,45. From these findings, the working diagnose for this patient was left posterolateral neck tumor. The boy then underwent an ultrasound examination. The result was a solid mass with minimal intratumor vascular.

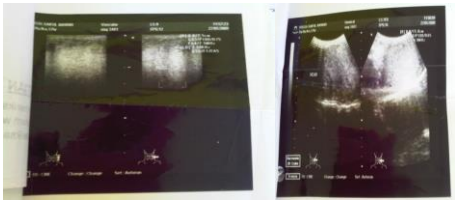

Figure 2. The ultrasound results of the mass in the left side of the neck

The consultation with the radiology department concluded that a head MRI was needed to determine whether the tumor was connected to the nervous system and bone. The results of the head MRI revealed a homogenous well defined mass, isotense on T1W1 and isohyperintense on T2W1 with heterogenous enhancement after contrast administration, on the left posterolateral neck, the size is average 11x17x23 cm. The mass was outside of the brain and cervical vertebrae. The cerebrum, ventricle and medulla spinalis within normal limit. The MRI conclusion was a left posterolateral embrional tumor colli outside of the brain and cervical vertebrae with differential diagnosis rhabdomyoma.

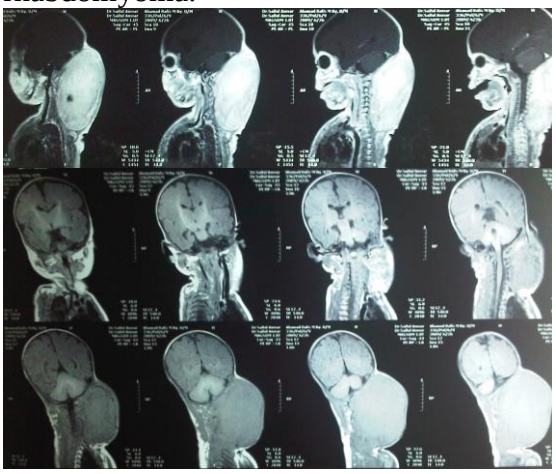

Figure 3. The MRI results

An incisional biopsy operation was planned. Macrossly was appeared a solid white mass. Under the microscope, the slice showed a tissue consisted of proliferation of spindle cell inside fybrocollagen tissue and stellate cell inside myxoid tissue. The biopsy concluded low grade fibromyxosarcoma. Therefore, a resection surgery and chemotherapy were planned. At the age of 3 months, the boy underwent a complete resection of the mass. The mass was solid and weighed 500 grams.

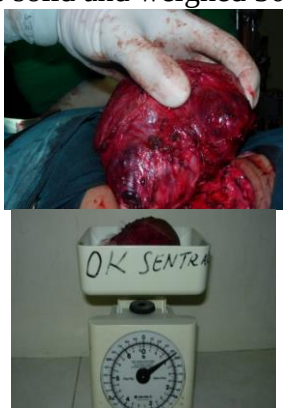

Figure 4. The resected mass

After the operation, the boy underwent chemotherapy course for fibrosarcoma consisting of cyclophosphamide, adryamicin,vincristine and DTIC. After completing this full course for 4 months, the patient underwent an evaluation by MRI. The MRI showed no residual mass.

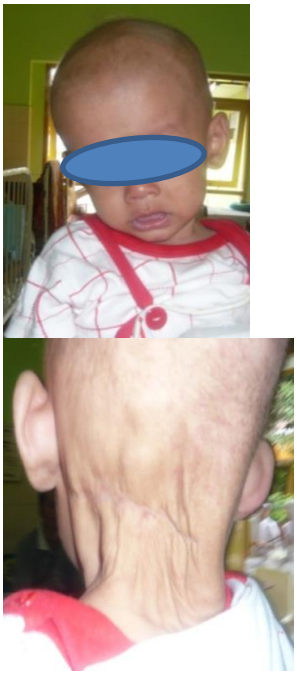

Figure 5. Left: The boy after full course chemotherapy. Right: Scar tissue after surgery.

DISCUSSION

Fibrosarcoma is a mesenchymal tumor of adult and pediatric age groups representing 10 to 12% of all soft-tissue sarcomas. It represents less than 1% of all childhood cancers but is the most common soft-tissue sarcoma in those under 1 year of age. In more than one-third of patients, it is present at birth and a second peak of incidence occurs between 10 and 15 years of life. The tumor may be observed at birth, in infancy or in childhood, and has been called congenital or infantile fibrosarcoma. 75% in children less than 10 years old including 36% in newborns (congenital fibrosarcoma). Ratio of boys to girls is 1.2 : 1, it is locally aggressive with a rare metastatic spread and a good rate of survival. The location of fibrosarcoma in children, ranked by frequency is in the lower extremities, upper extremities, head and neck, trunk, pelvic area but rarely the retroperitoneal or visceral area and chest. [1,9]

In this case, the patient had mass in the neck since birth with a slight enlargement of the mass. No history of fever, decreased of body weight or bleeding, and there was no respiratory distress nor difficulty of feeding.

Fibrosarcoma, like other soft-tissue sarcomas, has no definite cause. Current research indicates that many sarcomas are associated with genetic mutations and the more common genetic defects include allele loss, point mutations, and chromosome translocations However, no definite cause of fibrosarcoma is known, although genetic mutations may play a role. Several inherited syndromes are associated with sarcomas., for example, patients with multiple neurofibromas may have a 10% lifetime risk of developing a neurosarcoma or a fibrosarcoma.[10.11]

In this case, the cause of the tumor is still unknown and there was no history of malignancies in either his mother's and father's families.

The diagnostic evaluation for soft tissue tumors should delineate the extent of the primary tumor and the extent of metastatic disease and should consist of the following:

1. Complete history
2. Physical examination
3. Laboratory studies
4. Imaging studies : plain radiograph, Magnetic resonance imaging (MRI) and/or computed tomography (CT) of primary lesion, and ultrasonography (USG)
5. Biopsy , histopathological examination

Laboratory studies are generally not helpful during the initial evaluation, and plain radiographs of the involved anatomic region are needed to evaluate for primary or secondary bone involvement. For soft-tissue masses, size

often can be estimated, any bone involvement can be seen and intralesional content (matrix) can sometimes be determined. For bony lesions, plain radiographs often greatly assist in the diagnosis and the determination of location, size, and local extent of involvement. [9,10]

MRI is the best modality overall for examining soft-tissue masses and for detecting the intraosseous and extraosseous extent of many bony sarcomas. Also, MRI is useful in providing information about the local extent, lesion size, and involvement of the neurovascular structures In addition, MRI may have value in predicting the extent and probability of surgical resection for extremity, abdominopelvic, and retroperitoneal tumors because of its enhanced ability to differentiate tumors from normal surrounding tissue compared to CT Scan. Furthermore, it is useful for evaluating spinal cord involvement with retroperitoneal or paraspinal tumors. Although imaging findings are not specific of infantile fibrosarcoma, diagnosis can indicated when MR imaging depicts a large well-circumscribed mass arising in a limb at birth or during the neonatal period. This mass is sometimes heterogeneous and septate and exhibits an isointense T1- and hyperintense T2-weighted signals and strong enhancement. MR is also the technique of choice for follow-ups during treatment which consist nowadays almost exclusively in chemotherapy. [10,11,12]

CT scanning is used to delineate bone involvement, bone destruction, or bone reaction especially for sarcomas arising in bone. The density of fibrosarcomas is similar to that of surrounding normal muscle. Signs of fracture or impending fracture may be seen, and the tumor can be more accurately localized. Some authors have suggested the use of gadolinium and ultrasound scans for diagnosis, but to date, the value of these tests for the staging of sarcomas remains limited. [10,11]

Ultimately, the diagnosis of fibrosarcoma is made with tissue obtained from a biopsy. Biopsies should be thought of as the first step towards treatment, rather than the last step in diagnosis. A biopsy should always follow a full radiographic workup and any biopsy performed must include an adequate volume of tissue. In centers with expert interpretation, core-needle biopsy or fine-needle aspiration may be acceptable The value of fine-needle aspiration is primarily to differentiate a solid mass from a fluid containing mass or an abscess for drainage. In all other instances, a minimum of a True-Cut needle biopsy is warranted.. An excisional biopsy of a lesion is undertaken only in those instances when the tumor is small (<2.5 cm) or situated so that an eventual wide local resection can be done without risk or functional deformity. In all other instances, an incisional biopsy is obtained and is carefully planned and placed, so that the tract can be completely excised at the time of definitive surgery.[5, 10]

Fibrosarcomas are tumors of malignant fibroblasts and collagen which vary in histologic grade. Infantile fibrosarcoma is histologically similar to those seen in adults. Fibrosarcoma occurred mainly in the muscles of the extremities . This congenital form shows uniform fibroblasts or myofibroblast with low rate of mitosis. [9,10]

The tumor shows a biphasic pattern with fi brous and myxoid areas, minimal nuclear pleomorphism, low to moderate cellularity, and a swirling, whorled growth (A: HE, ×200). The background matrix ranges from fibromyxoid to densely fibrous (B: HE, ×400)

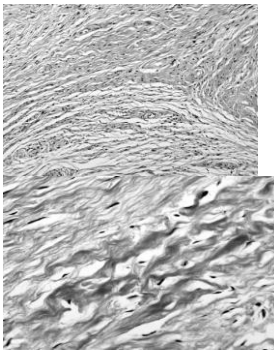

Figure 6. Histopathological slice of fibrosarcoma

There were some differential diagnosis for fibrosarcoma :

- Nodular fascitis
- Myositis ossificans
- Inflammatory pseudotumor
- Neurofibrosarcoma
- Malignant peripheral nerve sheath tumor/schwannoma
- Poorly differentiated embryonal rhabdomyosarcoma
- Monophasic (spindle cell) synovial sarcoma

Many parameters are helpful in the differential diagnosis of infantile fibrosarcoma. However, the number of inflammatory cells, histologic pattern and age of the patient is helpful in differential diagnosis of infantile fibrosarcoma [2,5,8,9,10]

In this case, the diagnosis of infantile fibrosarcoma was established from history taking, physical examinations, imaging and also a biopsy. The mass in the left neck was palpated as a firm, non-tender and immobile mass, the overlying skin was tight and within normal color. The mass size was about 17 cms in diameter and there were no other palpable masses in other parts of his body. The ultrasound examination result showed a solid mass with minimal intratumor vascular, followed by a head MRI with conclusion: a left posterolateral embrional tumor colli outside the brain and cervical vertebrae, differential diagnosed as rhabdomyoma. An incisional biopsy operation was planned. And the histopathological examination revealed a low grade fibromyxosarcoma.

Nonrhabdomyosarcomatous soft tissue sarcomas (NRSTS) occurring in children are a diverse group of neoplasms whose rarity has hindered elucidation of the optimal treatment and prognosis. The general management of fibrosarcoma consists of surgery, chemotherapy and radiotherapy. Previously reported series of treatment outcomes in infantile fibrosarcoma have been limited to very few patients due to the rare occurrence of this tumor and initial chemotherapy combined with surgery has been successful in most cases. Unlike that for rhabdomyosarcoma, which is a highly chemosensitive tumor, the mainstay of treatment for NRSTS is complete surgical resection with or without adjuvant radiotherapy to prevent local recurrence.[1,14, 15]

Chemotherapy is more widely used in children than in adults because children better tolerate the acute adverse effects and, in general, have malignant diseases that are more responsive to chemotherapy than are malignant diseases of adults. In general, radiotherapy is used sparingly in children because they are more vulnerable than adults to its late adverse effects. [2]

Wide local excision is the primary form of treatment, but in some cases, as in our case, it is impossible to remove lesion because of the anatomic extent of these tumors without disrupting peripheral vessels and nerves. Sometimes, the adjacent bone also has to be excised and in larger series of patients with infantile fibrosarcoma, the primary and secondary amputation rate is approximately 50%. In irresectable tumors, in relapses and after incomplete surgery, chemotherapy, and in older

children, radiotherapy is an adjuvant therapeutic tool.[1,13]

The treatment choice of fibrosarcoma is still complete excision of the tumor but an initial chemotherapy can convert the surgical approach from mutilating approaches to more conservative surgery, or it can be curative for distant metastasis [14-16]

In this case, the patient underwent a resection surgery and chemotherapy. At the age of 3 months, the boy underwent a complete resection of the mass then followed by a 4 month regimen of chemotherapy that consisted of cyclophosphamide, adryamicin, vincristine and DTIC.

Infantile fibrosarcoma's clinical course is more favorable with a rare metastatic spread. The local recurrence rate is high, up to 43%, and recurrence may occur as late as 15 to 31 years after the initial operation. However, fibrosarcoma occurring in infants has an excellent outcome despite histological findings. Initial complete excision should be attempted in all children and recurrence indicates a more aggressive variant and warrants more aggressive complete re-excision wherever possible. The overall disease-specific survival for children with NRSTS of the head and neck at M. D. Anderson was 80% at 2 years and 75% at 5 years[1,16, 17,18]

Prognostic factors in children with NRSTS include the presence or absence of etastatic disease, surgical respectability of the lesion, histological grade, tumor invasiveness, and size of the lesion. For congenital fibrosarcoma in children, the prognosis (which is related to age and to time of diagnosis) is much better, with the disease having long-term survival rates of higher than 50%.[5, 10, 20]

As with all sarcomas of the musculoskeletal system, successful treatment of fibrosarcoma must be accompanied by an organized plan for clinical follow-up. This often involves a schedule of repeat examinations and diagnostic studies. Patients often are monitored for a minimum of 5 years and at preset intervals, the patient is reexamined, and plain radiographs of the involved site are obtained. Repeat staging studies of the local area and of the chest also are performed. [10]

In this patient, the boy had completed the chemotherapy course. The patient underwent an evaluation by MRI. The MRI showed no residual mass. The boy still control routinely to the pediatric department.

SUMMARY

A rare case of infantile low grade fibromyxosarcoma had been reported. Based on the history, physical examination, laboratory examination, and radiologic imaging, the diagnosis of infantile low grade fibromyxosarcoma was established. MRI is the best modality overall for examining soft-tissue masses and is useful in providing information about the local extent, lesion size, and involvement of the neurovascular structures. In addition, MRI may have value in predicting the extent and probability of surgical resection for these tumors compared to CT Scan. Furthermore, it is useful for evaluating spinal cord involvement with retroperitoneal or paraspinal tumors.

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