

INFANTILE FIBROSARCOMA PRESENTING AS A BLEEDING, CONGENITAL NECK MASS

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Abstract

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Background: Infantile fibrosarcoma represents the most common soft tissue sarcoma in children under 1 year age, but constitutes less than 1% of all childhood tumors.

Case presentation: We report a case of congenital infantile fibrosarcoma in a newborn, which came to us with an unusual presentation of profusely bleeding neck mass, mimicking a hemangioma and was subsequently diagnosed and adequately treated by wide local excision.

Conclusion: Very high suspicion must be kept for any firm to hard mass in the extremities or trunk and neck. Proper surgical planning and wide local excision is the treatment of choice and when required chemotherapy must be administered for functional preservation

Introduction

Fibrosarcoma constitutes 10-12% of all soft tissue sarcomas in adult and pediatric age-groups, representing less than 1% of all tumors in childhood. It is the most common soft tissue sarcoma in infants. The tumor may manifest at birth, in infancy or in later childhood. It has a different behavioral pattern than its adult counterpart. It may be locally aggressive, but rarely metastasizes and has a good overall prognosis after treatment by adequate surgical resection.

Case report

A full-term new born male child presented to us with a mass in the left side of neck. The child was a product of non-consanguineous marriage and delivered by caesarean section due to fetal distress. At birth a red, violaceous, pedunculated mass was noted on the left side of neck and the child was referred to us further care with a preliminary diagnosis of hemangioma neck. During transportation, the mass bled profusely due to trauma by baby's own hand, which further added to the diagnostic dilemma.

On examination the baby weighed 2800gms and was hemodynamically stable. On local examination, the mass located in the anterior triangle of the neck on the left side was pedunculated, lobulated with a narrow stalk. It was 4 x 4cm size, red-violaceous in colour with prominent vascular markings, firm to hard, with ulcerated surface. No bruits could be heard over the swelling and there was no local lymphadenopathy. Systemic examination was within normal limits. Complete blood counts with coagulation profile and biochemical tests were within normal range. In view of the constant oozing post-trauma, wide local excision of the mass was done with 1 cm margin all around. On cut section, it was homogeneously grayish white in colour, without any areas of necrosis or hemorrhage. On subsequent pathological examination on light microscopy, superficial portion of the lesion was found to be comprised of long and short, intersecting fascicles of spindle cells with bland nuclear chromatin. The deeper portion

showed a varied growth pattern with a high mitotic rate and numerous ectatic and slit-like 'staghorn' vessels. There was deposition of intercellular collagen. All features were suggestive of infantile fibrosarcoma.

The child is now 3 months in follow up and there is no evidence of recurrence or metastasis. He is planned for a long term follow-up.

Discussion

Fibrosarcoma is a spindle cell tumor of mesenchymal origin, representing approximately 10% of all soft tissue sarcomas. Though it is rarely found in the pediatric age group accounting for less than 1% of all childhood malignancies, it is the most common soft tissue sarcoma in patients less than 1 year of age.⁽¹⁾ More than one third patients present at birth and a second peak occurs between 10-15 years of life. It is called true congenital fibrosarcoma if it occurs before 3 months age. The remaining pediatric fibrosarcomas are called infantile or juvenile.⁽²⁾ Histologically, infantile fibrosarcoma (IFS) has a relatively similar picture as adult fibrosarcoma, but it has more benign course than its adult counterpart and the lesions detected during infancy have the best prognosis. This difference in biological behavior qualifies infantile fibrosarcoma as a distinct entity.⁽³⁾

Clinical features

The most common site of involvement of IFS is distal part of extremities like foot, ankle, hand, wrist, fore arm followed by trunk, head and neck.⁽²⁾ The clinical presentation is usually of a nontender, exophytic mass that may be as large as 20 cm. The mass grows steadily and may enlarge rapidly. Ulceration of the overlying skin due to intra-lesional bleeding may occur. Occasionally these masses are more vascular, mimicking hemangiomas clinically and may sometimes bleed profusely if injured.⁽⁴⁾

The mass is locally aggressive with the incidence of metastasis being less than 10% in patients younger than 5 years of age but rising above 50% beyond 10 years.⁽¹⁾ Local recurrence rate is 20-43%, occurring mostly within the first 12 months of initial surgical resection.⁽⁵⁾ The overall survival rate of IFS has been reported to be in the range of 90 to 100%, with a greater risk of mortality in patients with truncal lesions. There are few reports in literature of spontaneous regression of untreated or incompletely resected fibrosarcomas.

Pathology



Figure 1.A Infantile fibrosarcoma of neck mimicking a hemangioma; pre-operative



Figure 1.B Immediate post-operative

IFS is classified as a low grade non-rhabdomyosarcoma soft tissue sarcoma (NRSTS) and according to the pediatric oncology group classification of NRSTS, it is classified as grade I. Histologically, oval to spindle shaped cells with hyperchromatic nuclei, showing little variation in size and shape are identified (Figure 1). A fascicular, 'herringbone' pattern is often present. IFS have greater cellularity, mitotic activity and hyperchromatism than fibromatosis, but differentiation between the two may sometimes be difficult (Figure 2).⁽⁶⁾

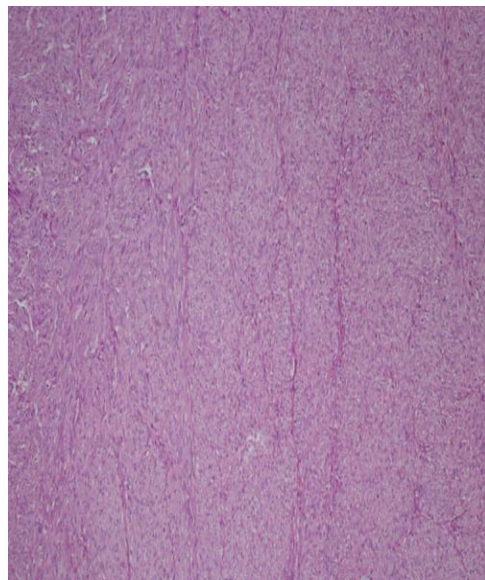


Figure 2.A (X200 magnification) Interlacing fascicles of spindle shaped tumor cells

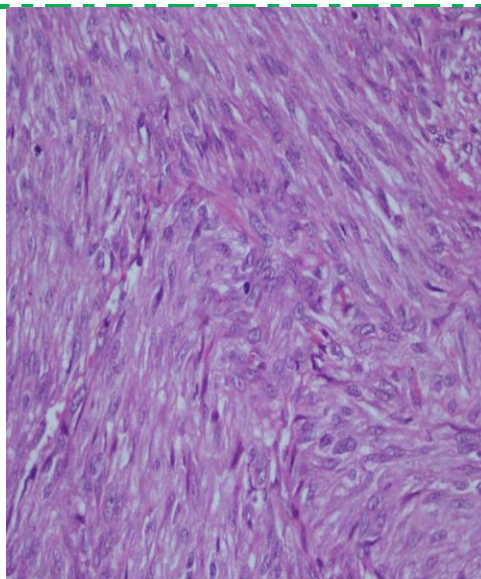


Figure 2.B (X400 magnification) Pleomorphic tumor cells with atypical mitotic figures

Recently, immunohistochemical methods are being used for confirmed diagnosis of IFS. Fibrosarcoma shows lack of staining for desmin, S-100 protein and factor VIII and shows cytoplasmic staining for vimentin.

Molecular markers and genetics

Non-random gains of chromosome 8, 11, 17 and 20 are characteristic molecular marker of IFS but not for adult fibrosarcoma, noncellular fibromatosis or myofibromatosis. A chromosomal translocation [t(12:15)(p13;q25)] typical to IFS has been detected, which gives rise to ETV6-NTRK3 gene fusion. This is useful in distinguishing congenital fibrosarcoma from other malignant and bony spindle cell tumors.

Differential diagnosis

The differential diagnosis of a soft tissue mass in children includes hemangioma, lymphangioma, lipoma, hemangiopericytoma, rhabdomyosarcoma, myofibromatosis among others.⁽⁷⁾

Radiology

Radiological appearances of IFS are non-specific, though the detection of a well-demarcated soft tissue tumour in an infant should strongly suggest its diagnosis. Ultrasonography is used for initial assessment. CT is useful in demonstrating the extent of tumor, especially when there is bony involvement. MRI is the imaging modality of choice for evaluating the nature and extent of this soft tissue tumor and its relationship to surrounding structures. It is important for planning surgery and monitoring the chemotherapeutic response and also for early detection of recurrence. On MRI, IFS appear as a well demarcated isointense lesion on T1 weighted images and hyperintense on T2 weighted images. Areas of hypointensity may be seen on T2 weighted images, which represent hemorrhage or areas of myxoid degeneration. MRI is useful in differentiating IFS from hemangioma as the latter appears as a well-defined mass with homogenous rather than heterogenous enhancement.

Biopsy

A properly planned incisional biopsy is the procedure of choice for definite diagnosis.⁽⁵⁾

Management

The standard treatment modality for IFS is surgical excision. A simple excision biopsy is inadequate for therapy and may result in incomplete resection and future recurrence. Wide local excision encompassing histologically tumor free margins should be the primary form of treatment.⁽⁵⁾ At least 1 cm of tissue beyond the obvious lesion should be removed. Whenever possible, the excision should be performed without sacrificing any significant function of the neighbouring structures.⁽¹⁾

The role of chemotherapy as primary treatment of IFS remains unproven. In cases of non-resectable tumors, incomplete surgical excision or relapses, chemotherapy definitely is an adjuvant therapeutic mode. There are encouraging results with chemotherapy in fibrosarcoma involving the extremities and with proper chemotherapy, extremity sparing surgery is also possible. The most common chemotherapy regimen consists of vincristine, actinomycin-D and cyclophosphamide (VAC). In few cases, ifosfamide and doxorubicin (adriamycin) may be substituted for cyclophosphamide and actinomycin-D respectively.⁽⁸⁾

The role of radiotherapy in the treatment of IFS remains controversial. Distant metastasis can be treated with resection or chemotherapy.

Conclusion

Though infantile fibrosarcoma is the most common soft tissue sarcoma in the newborn and infancy, it as such is very rare but serious tumour. Very high suspicion must be kept for any firm to hard mass in the extremities or trunk and neck, as the presentation can be varied. Proper surgical planning and wide local excision must be done and when required chemotherapy must be administered for functional preservation.

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REFERENCES

1. Soule EH, Prtichard DJ. *Fibrosarcoma in infants and children: A review of 110 cases. Cancer* 1977;40:1711-1721
2. Chung EB, Enzinger FM. *Infantile fibrosarcoma. Cancer* 1976;38:729-739
3. Cofer BR, Vescio PJ, Wiener ES. *Infantile fibrosarcoma: Complete excision is the appropriate treatment. Ann Surg Oncol* 1996;3:159-161
4. Boon LM, Fishman SJ, Lund DP, Mullikan JB. *Congenital fibrosarcoma masquerading as congenital hemangioma: Report of two cases. J Pediatr Surg* 1995;30:1378-1381.
5. Trobs RB, 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: 123-128.
6. Madden NP, Spicer RD, Allibone EB, Lewis IJ. *Spontaneous regression of neonatal fibrosarcoma. Br J Cancer* 1992;66:suppl XVIII,S72-S75
7. Akyuz C, Sari N, Vargel I, Gedikoglu G, Haliloglu M, Buyukpamukcu M. *A new born with infantile fibrosarcoma of foot: Treatment with chemotherapy and extreme-sparing surgery. J Perinatol* 2010;30: 63-65..