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Case Report

Infantile Fibrosarcoma Presenting as Neck Mass: A Case Report

Author
Dr Yasap Tana
Consultant Pathologist, Department of Pathology, Tomo Riba Institute of Health and Medical Sciences, Naharlagun, Arunachal Pradesh, India
(Former Senior Resident, Department of Pathology, Government Medical College, Thrissur, Kerala, India)
*Corresponding Author
Dr. Yasap Tana

Abstract
A one year old boy presented with a mass in the posterior triangle of left side of neck for ten months with rapid increase in size since one month. Radio-imaging was done. He underwent surgery, the mass was excised and the histopathological examination was done. Clinically, it was thought to be as schwannoma. Tumor measured 10 x 8 x 6 cm approximately, fixed with underlying tissue, venous prominence seen on bosselated surface and was non-tender.
Microscopically, showed a cellular neoplasm composed of spindle cells arranged in herringbone pattern, also as fascicles and bundles, with focal area showing slit like thin-walled vessels simulating hemangiopericytoma like vasculature. Focally, shows infiltration of muscle and fat. Immunohistochemistry study was performed.
Correlating histology with clinical, radiology and immunohistochemistry findings, the patient was diagnosed as a case of Infantile Fibrosarcoma of neck region, a relatively rare malignant fibroblastic tumor, occurring mostly in infancy, often misdiagnosed with benign tumor.
Keywords: *Cellular, herringbone pattern, hemangiopericytoma, immunohistochemistry, fibroblastic.*

Introduction

Infantile Fibrosarcoma (IFS), also known as congenital fibrosarcoma, a relatively rare low grade but locally aggressive malignant spindle cell or fibroblastic tumor of mesenchymal origin**.** It includes approximately 10% of all sarcomas in children[1].

The highest prevalence of IFS is in early in life, with over 75% of cases recorded in the first year of life and 15% in the second year of life[2,3]. It is mostly presented as a tumor in extremities, trunk, followed by head and neck but can occur anywhere in the body.

Histologically, it is similar to adult-type but with a better prognosis. Although recurrence of infantile fibrosarcoma is common, metastatic spread is infrequent[1]. It has a greater chance of long-term survival (90% at 5 years) and more chemosensi-tivity than the adult fibrosarcoma[4].

Surgery is considered the main choice of treatment. The use of adjuvant therapy and chemotherapy is not clear yet, but mostly used for high grade tumors

with microscopic metastasis[5] .

It is generally misdiagnosed because of histological similarities to benign tumors. Ultimate diagnosis is made by physical examination, special radiologic studies and biopsy.

Case Report

A one year old boy presented with swelling in left posterior triangle of neck for ten months, gradually increasing in size with history of recent rapid increase since one month. Clinically, it was thought to be as Schwannoma.

On local examination, neck mass measured 10x8x6 cm, fixed with underlying tissue, non-tender, bosselated surface showing venous prominence (Fig.1).

The contrast enhancing computed tomography (CECT) revealed a well-defined heterogenously enhancing soft tissue density in the posterior triangle of left side of neck (Fig.2).

The patient underwent surgery, the mass was excised and histopathological examination of the specimen was done.

On gross, received an irregular nodular mass measuring 7x6.5x6 cm, weighing 75 gram. (Fig.3, Fig. 4). Outer surface was painted with black ink. Cut surface of the mass showed a fairly circumscribed, homogenous, grey-white, firm growth with somewhat pushing borders. On serial sectioning, appearance was similar. (Fig.5). Multiple bits were taken from multiple areas including entire circumference of tumor mass.

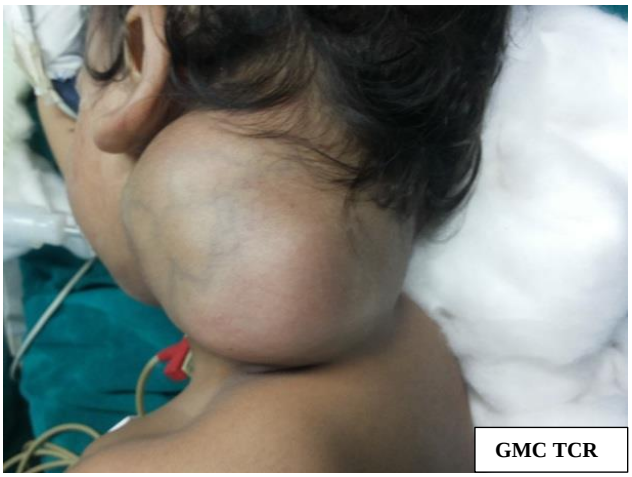

Figure.1 Mass in left side of neck with engorged veins on the surface

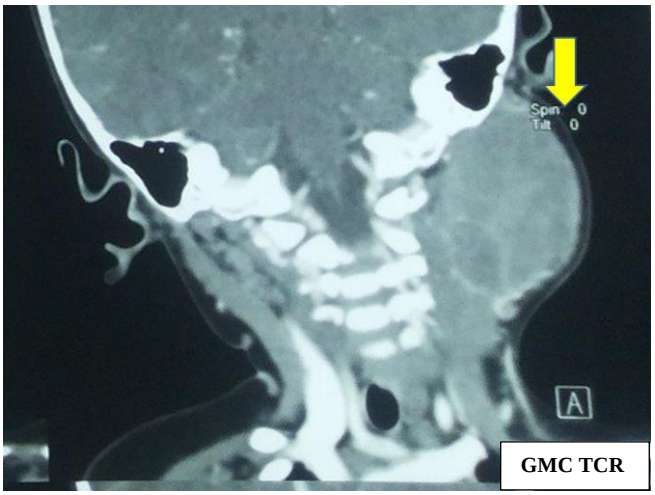

Figure.2. CECT scan image well-defined heterogenously enhancing soft tissue density in the posterior triangle of left side of neck.

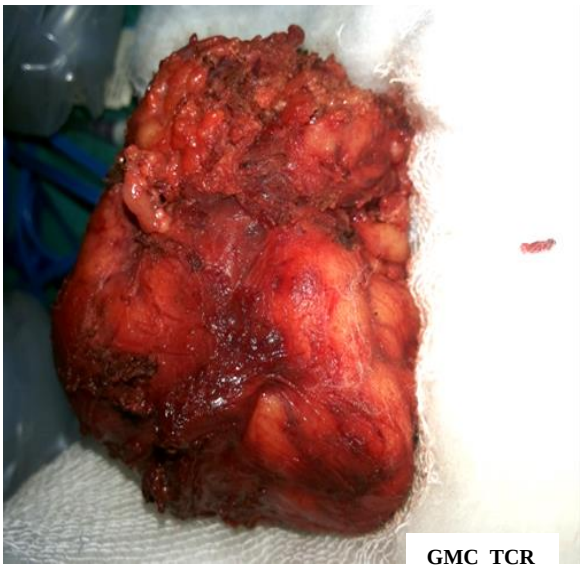

Figure.3 Excised mass immediately after excision.

Figure.4 Outer surface of mass painted with black ink.

Figure.5 Cut surface of the mass showing homogenous, grey-white, firm with pushing borders.

On microscopy, Hematoxylin and Eosin (H&E) stained sections showed cellular neoplasm composed of cells arranged in herringbone pattern, also seen arranged in fascicles and bundles, with focal area showing slit like thin-walled vessels simulating hemangiopericytoma like vasculature. A foci showing infiltration of muscle and fat by tumor cells seen. Individual cells have moderate amount of cytoplasm with elongated, ovoid to round vesicular nuclei, some cells showing prominent nucleoli. Mitotic activity seen i.e, 4 to 5 mitoses per high power field, with occasional atypical mitosis. (Fig.6 to Fig.12)

Grossly and microscopically no lymph nodes or adjacent tissue to the mass were identified to comment on margin status.

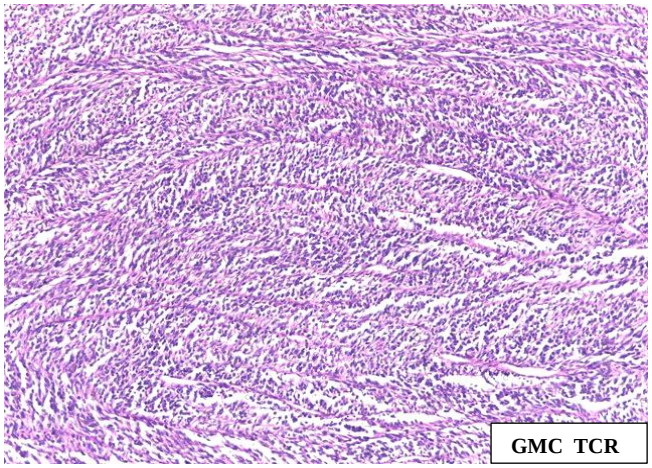

Figure.6 Arranged in herringbone pattern. (H&E, 10X magnification)

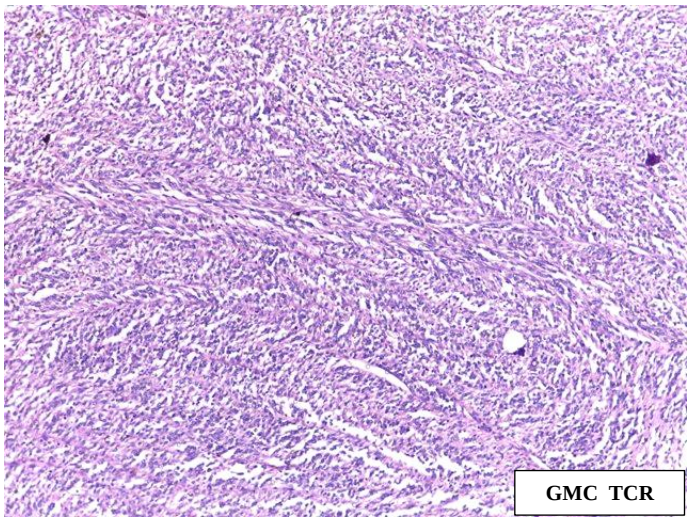

Figure.7 Uniform, well-oriented fibroblasts arranged in fascicular growth pattern. (H&E, 10X magnification)

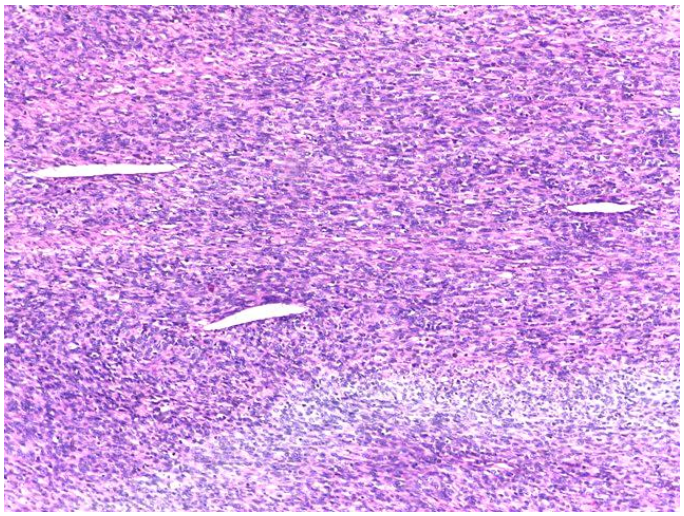

Figure.8 Hemangiopericytoma-like vascular pattern (H&E, 10X magnification)

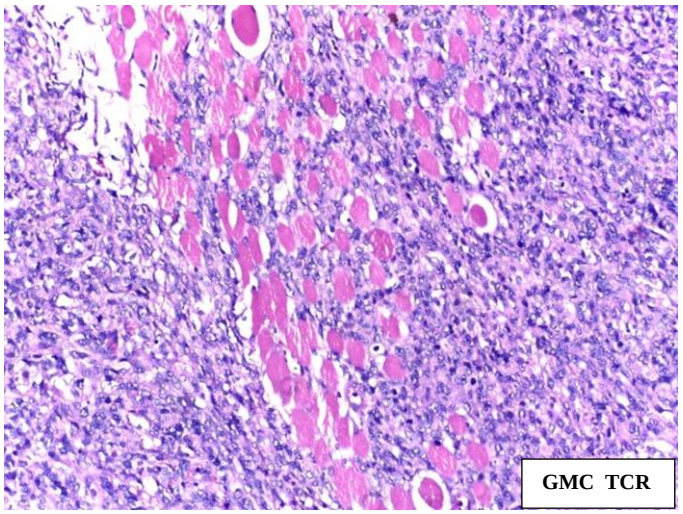

Figure.9 An area showing infiltration of muscle by tumor cells. (H&E)

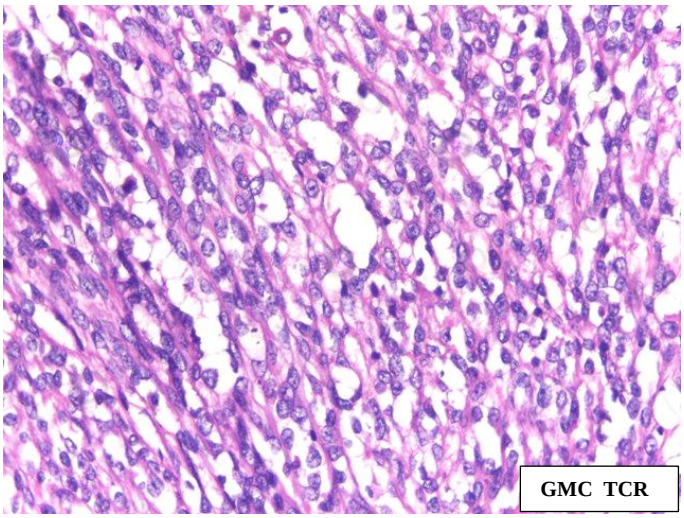

Figure.10 The cells show moderate amount of cytoplasm with ovoid to round vesicular nuclei showing prominent nucleoli. (H&E, 40X magnification)

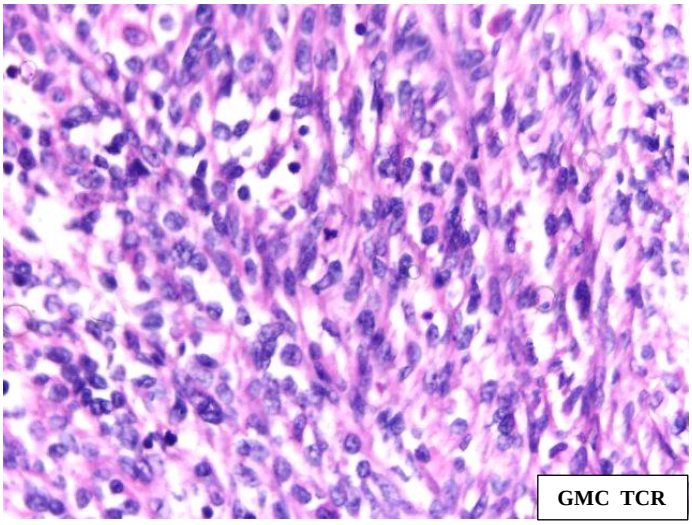

Figure.11 Atypical mitosis (H&E, 40X magnification).

Hence, the differential diagnoses based on histomorphology were infantile fibrosarcoma, Infantile Fibromatosis, Infantile Hemangiopericytoma, Spindle cell Rhabdomyosarcoma and monophasic Synovial Sarcoma.

Table.1 Differentiating features amongst close differential diagnoses.

FEATURES	INFANTILE FIBROSARCOMA	INFANTILE FIBROMATOSIS	INFANTILE HEMANGIOPERICYT-OMA
Gross	Subcutaneous	Poorly circumscribed, deep-seated	Nodule
Pattern	Herringbone, fascicular	Diffuse / streaks	Fascicles/whorls
Nuclear atypia	Present	Absent	Absent
Mitoses	Few to many	Rare	Infrequent
Necrosis	Frequent	Absent	Unusual

Immunohistochemistry (IHC) study was performed with a panel of markers i.e, Epithelial membrane antigen (EMA), Cytokeratin (CK), Desmin, Myogenin, CD 34, S100 and Smooth muscle actin (SMA), thereby narrowing down the number of differential diagnoses. As such there is no specific IHC marker for IFS. In this case, IHC study favored the possibility of a soft tissue neoplasm with myogenic and/or myofibroblastic differentiation. Meanwhile, Pan-Trk antibody was less known and unavailable at our centre.

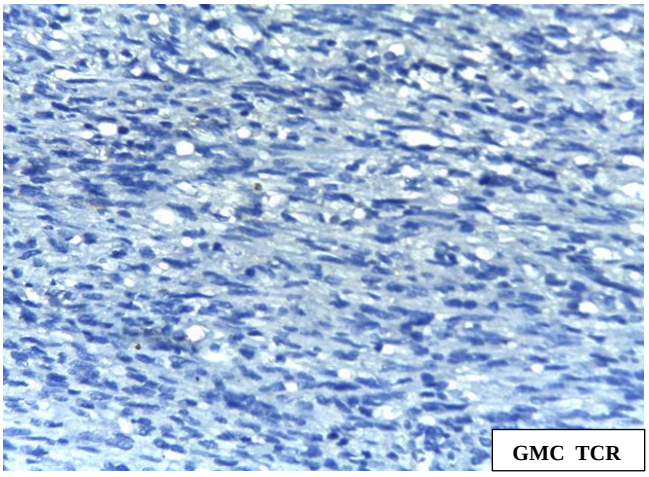

Figure.12 Histopathology: immunohistochemistry examination with EMA (40X magnification) showing negative expression in tumor cells.

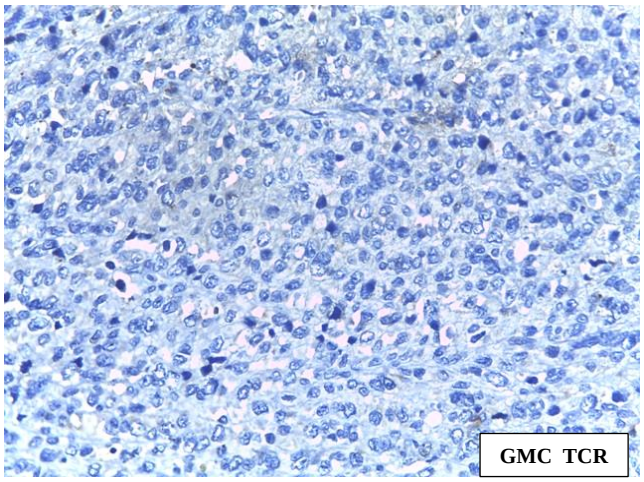

Figure.13 Histopathology: immunohistochemistry examination with CK (40X magnification) showing negative expression in tumor cells.

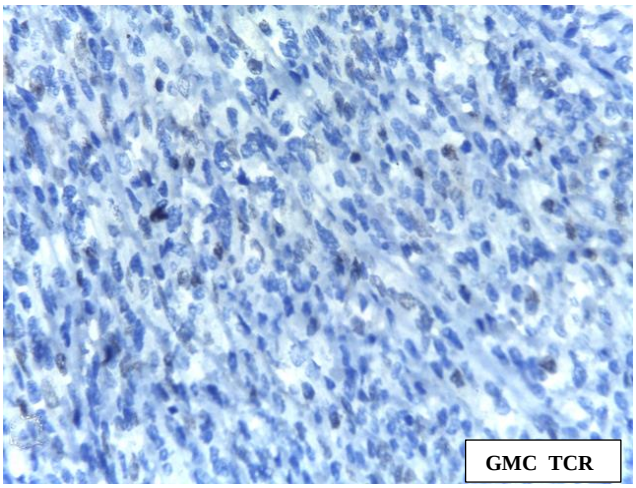

Figure.14 Histopathology: immunohistochemistry examination with Myogenin (40X magnification) showing negative expression by tumor cells.

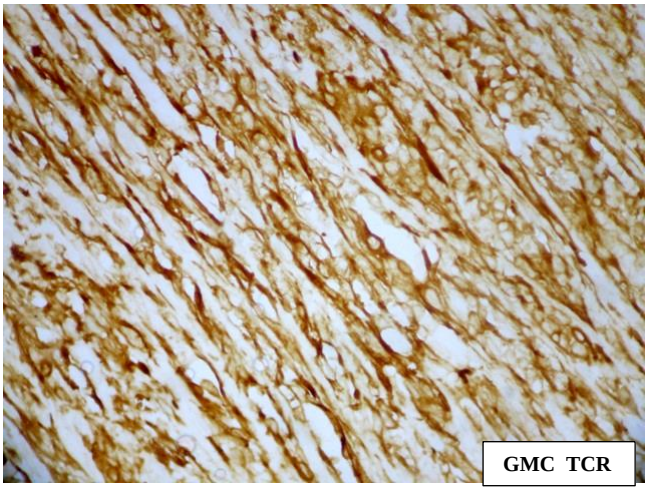

Figure.15 Histopathology: immunohistochemistry examination with Desmin (40X magnification) showing cytoplasmic positivity in tumor cells.

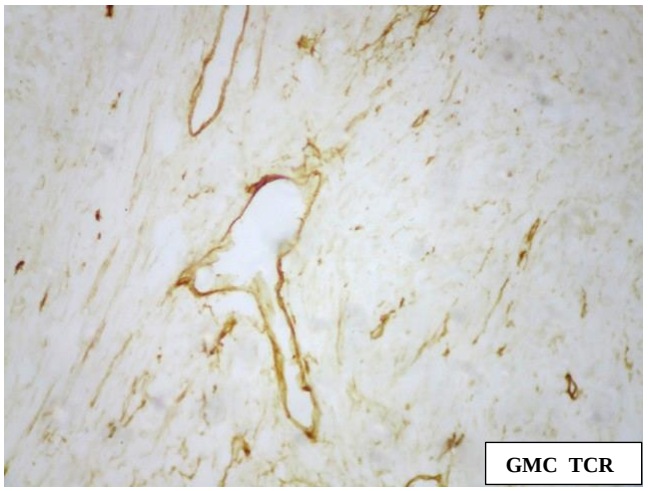

Figure.16 Histopathology: immunohistochemistry examination with CD34 (40X magnification) showing negative expression in tumor cells.

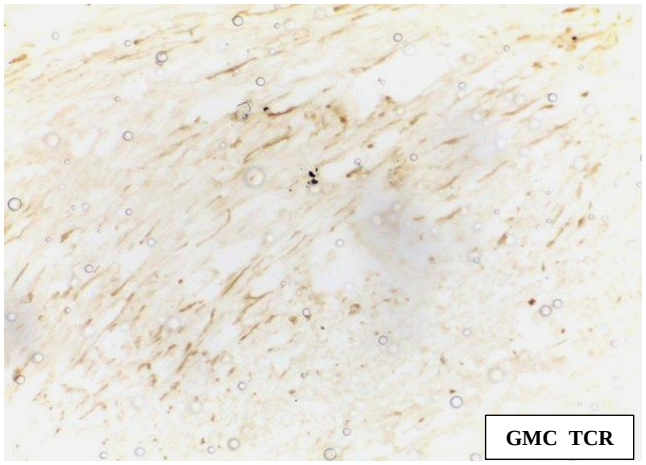

Figure.17 Histopathology: immunohistochemistry examination with S-100 (40X magnification) showing negative expression in tumor cells.

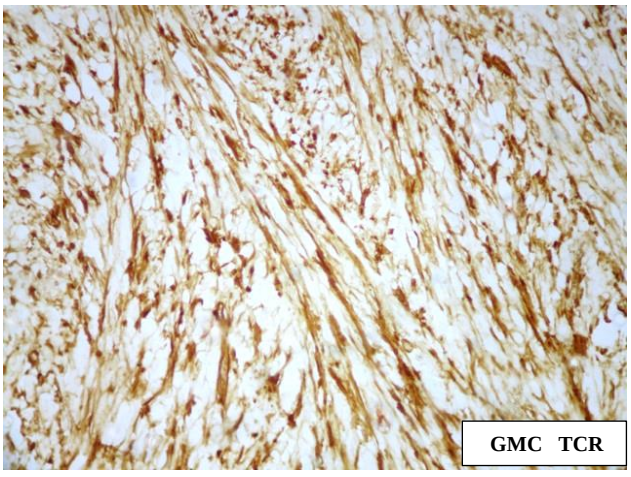

Figure.18 Histopathology: immunohistochemistry examination with SMA (40X magnification) showing cytoplasmic expression in tumor cells (tram-track like).

Correlating histomorphology with clinical, radiological and immunochemistry findings, final diagnosis of the mass lesion was made as Infantile fibrosarcoma.

In this case, unfortunately, patient could not be followed up.

Discussion

IFS is a rare malignant soft tissue tumour in the paediatric population accounting for 5-10% of all sarcomas in infants less than 1 year of age[6] . It is more prevalent in boys (60%) and primarily affects the lower and upper distal extremities (72%), followed by the trunk and the head and neck. Clinically, the most frequent manifestation of IFS is a distal, growing mass with no distinct limits[7] .

The tendency of IFS to mimic other tumors may interfere with the correct diagnosis, clinico-radiologically, often diagnosed as hemangioma and is believed to be one of the reasons for its rarity. When compared with adult fibrosarcoma, the clinical course of IFS is favourable. Most studies have found that cellularity, mitotic counts and extent of tumor necrosis do not correlate well with clinical behavior [8, 9].

On IHC, the spindle cells of IFS stain variably for muscle markers, including muscle-specific actin (MSA), smooth muscle actin (SMA), and h-caldesmon[10,11]. The more primitive-appearing ovoid cells tend not to express these muscle markers.

As per recent cytogenetic and molecular studies, NTRK3 aberrations are characteristic of this tumor and a Pan-Trk antibody was recently found to correlate well with the presence of NTRK3 fusions[12,13] .

RT-PCR (real-time polymerase chain reaction) and FISH (fluorescent in situ hybridization) demonstrated a unique reciprocal translocation, t(12;15) (p13;q25), resulting in ETV6/NTRK3 gene fusion in IFS. Via the adaptor IRS-1, the ETV6/NTRK3 protein is involved with many signalling cascades, including Ras-MAP kinase and PI3K-AKT. As a result, this translocation becomes a pathognomonic feature of IFS[14] .

Despite rapid growth and a high degree of cellularity, most are cured by wide local excision. It is reported that preoperative chemotherapy is useful for decreasing tumor bulk, enabling a more conservative surgical approach[15]. Thus, adjuvant radiotherapy and chemotherapy should be reserved for those IFS cases that are unresectable or have recurred or metastasized. The probability of a local recurrence is as high as 43% mostly due to incomplete resection[16]. The overall 10-year survival rate with standard treatment regimens (various combinations of surgery and/or standard chemotherapy) is about 90% [17] .

Lymphadenectomy is usually unnecessary since IFS seldom metastasizes to lymph nodes and there was no lymph node enlargement in our patient too.

Radiotherapy is typically not recommended as it can inhibit growth in child; it is only appropriate for axial primary sites in case total resection is impossible[5] .

Conclusion (s)

IFS is an uncommon tumor in early childhood with an overall favourable outcome than adult fibrosarcoma. It is locally aggressive but with relatively infrequent metastases and recurrence mostly in incompletely resected cases. Wide local excision is the initial surgical procedure of choice. It is indeed a diagnosis of exclusion and should stain negative for S-100, EMA and CK while excluding mimickers. Patients should be followed up for detecting further relapse or metastasis especially in older ages.

Department and Institution Where Work Was Done

Department of Surgery and Department of Pathology, Government Medical College, Thrissur, Kerala, India.

Declaration of Figures' Authenticity

All figures submitted are original, created by the author with no duplication and have not been previously published in whole or in part.

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