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INFLAMMATORY FIBROSARCOMA ON THE RIGHT LEG OF AN INFANT-A RARE CASE REPORT

Paediatric Surgery

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ABSTRACT

Infantile fibrosarcoma (IFS) is a rare tumor. The highest prevalence is in the first 2 years of life. Cases typically manifest in the extremities, with rapid initial growth, are non-tender, and are a poorly delimited masses that vary in size or consistency and lack sensitivity. The treatment for IFS is a multimodal approach, and surgical excision has become the main treatment. This report aims to present the results of our work on IFS cases. **Case Report:** A 7-month-old malewas admitted to our hospital with a chief concern of a mass in the right lower leg that first appeared 3 months ago. Physical examination showed a single mass with dense-rubbery consistency, tenderness, and limited range of motion.. MRI examination revealed isointense lesion on T1W and heterogeneously hyperintense on the T2W/STIR sequence measuring 5.4(tr) x 5.2(ap) x 5.6(cc) cm.Biopsyshowed spindle cell tumor, morphology favors infantile fibrosarcoma which was consistent with IFS. A local excision was made to assess for pathological anatomy. **Conclusions:** Infantile fibrosarcoma (IFS) is an uncommon tumor in early childhood. It tends to mimic other tumors, which may interfere with the correct diagnosis, which may contribute to its rarity. Local excision is the first treatment choice, with a good prognosis for IFS.

KEYWORDS

INTRODUCTION:-

Myofibroblastic and fibroblastic tumors in children are a quite prevalent subset of soft-tissue proliferations that exhibit a variety of clinical symptoms. Based on their biological behavior, these tumors have been classified as malignant (fibromyxoid sarcoma, adult fibrosarcoma), intermediate–locally aggressive (lipofibromatosis, desmoid fibroma), intermediate–rarely metastasizing (inflammatory myofibroblastictumors, infantile fibrosarcoma, low-grade myofibroblastic sarcoma), and benign (myositis ossificans, myofibroma, fibromatosis colli).[1,2]

Soft-tissue sarcomas (STS) comprise around 8% of all pediatric malignancies; they are commonly classified as either rhabdomyosarcoma (RMS) or non-RMS-STS. A non-RMS-STS that affects both adults and children, fibrosarcoma frequently has a better prognosis in younger patients [3]. Approximately half of IFS occurrences take place in the limbs.

Over 75% of instances of the rare tumor inflammatory fibrosarcoma (IFS) are reported in the first year of birth, and 15% are reported in the second year [2, 3]. IFS is most common early in life. Prenatal ultrasonography screening has already found less than 15% of cases [4].

It usually presents as an insensitive, non-tender, poorly defined mass that fluctuates in size and consistency, with fast initial growth occasionally followed by a more indolent course. Compared to the adult version of fibrosarcoma, IFS has a higher chance of long-term survival (90% at 5 years), a lower incidence of metastasis (10%), and a higher degree of chemosensitivity.[4,5]

In 48–62% of instances, primary tumors are determined to be incurable at diagnosis, requiring a multimodal strategy that includes local therapy, conservative surgery, and preoperative cytoreductive chemotherapy. Surgical excision has been the primary course of treatment in recent decades. Some illnesses may require surgical treatment in addition to chemotherapy and radiotherapy since primary excision may involve an intrusive, potentially painful surgery with a greater morbidity [6,7]. This report describes our experience treating an IFS case because it is uncommon.

Case Report: -

A 7-month-old male, otherwise healthy baby, presented with swelling over his right leg above his ankle for 3 months. History of increase in the size of swelling present. The physical examination showed a 5.0cm x 5.0 cm mobile solid mass. Evidence of active bleeding is present from the swelling. No lymphadenopathy was present in the inguinal region and no generalized lymphadenopathy. There was no family history of malignant disease. The patient was advised MRI of the Right lower limb which revealed a large-sized lobulated infiltrative soft tissue mass lesion noticed at the posteromedial aspect of the leg above the ankle joint, seen infiltrating the underlying posteromedial muscular plane with a large exophytic component and with areas of necrotic changes.

Imaging Findings

The lesion measures 5.4(tr) x 5.2(ap) x 5.6(cc) cm,appears isointense on T1W and heterogeneously hyperintense on the T2W/STIR sequence. no fat component calcification or intralesional flow voids were noted.

Finding likely favored malignant neoplastic soft tissue lesion like Sarcoma.

Ultrasonography of the abdomen and pelvis did notdetect any other lesions.

Laboratory Findings

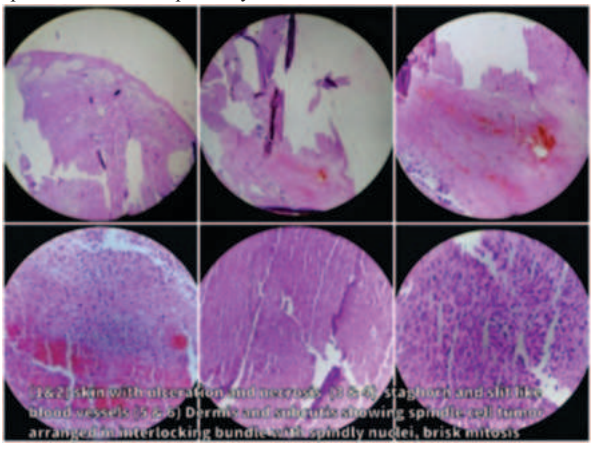
A blood test of complete blood count, kidney function test, and liver function test including alfa – fetoprotein were within normal limits.

Intraoperative Findings
A decision was taken to perform an excisional biopsy to conclude a definitive diagnosis. Intraoperatively mass was well defined and located in soft tissue immediately above the posteromedial muscle plane. The lesion was dissected out and removed completely macroscopically. The raw area that remained after the dissection was covered with a split-thickness skin graft taken from the medial aspect of the right thigh.

Histopathological Finding
The histopathological report was suggestive ofdermis and subcutis showing encapsulated spindle cell tumor arranged in interlocking bundle with spindly nuclei, brisk mitosis(2-3/hpf), staghorn and slit-like blood vessel, and moderate lymphocytic infiltrate. The impression was spindle cell tumor, morphology favors infantile fibrosarcoma.

Follow up
The patient came for a follow-up after 20 days with a complaint of reappearance of nodular structure at the previous operative site. Considering the recurrence of the tumor biopsy was taken.

Histopathological features are suggestive of a herringbone pattern of tumor cells, increased mitotic activity, and irregular infiltration into the surrounding tissue histological features are suggestive of malignant spindle cell sarcoma probably infantile fibrosarcoma.



Wide local excision was done and tissue was sent for histopathological examination suggestive of infantile fibrosarcoma margins free from tumor.

The patient was sent to a cancer speciality hospital for further management wherein whole-body FDG PET/CT scan was done showing no evidence of local recurrence or distant metastasis. NTRK Gene panel rearrangement assay results were positive for NTRK3 rearrangement showing the atypical signal pattern with loss of one green signal corresponding to 5' region of NTRK3 gene suggesting variant NTRK3 rearrangement.Given no residual disease and margins free from tumor patient was categorized into group II disease and observation was planned.

After one year follow up no recurrence was noted on repeat MRI scan.

DISCUSSION:
Mesenchymal cells, primarily spindle cell fibroblasts with uncontrolled growth, are the source of fibrosarcomas. Despite this, there is anvarying range in the histologic architecture of these samples; some may show an unstructured pattern of immature or even primitive mesenchymal cells, while others may have varied amounts of inflammatory cells mixed in with the mesenchymal cells [8]. According to reports, fibrosarcomas most frequently start in the tendons and deep fascial tissues, while they can also develop in the bone's periosteum or medullary canal [9]. Usually appearing in areas of connective tissue rich in collagen, fibrosarcoma grows rapidly over several weeks.

A deep-seatedtumor may remain undetected for a long time. When an invasive tumor compresses nearby tissue and organs, it can cause a variety of symptoms, such as pain, restricted movement, poor blood circulation, and micturition issues [8,9].

Infantile fibrosarcoma (IFS) typically affects the lower and upper distal extremities (72%) and is more common in males (60%) than in girls. Clinically, a distant, expanding mass without clearly defined boundaries is the most common symptom of IFS. On rare occasions, the histology shows a structure like a hemangiopericytoma, and the surface is necrotic or ulcerative, resembling vascular malformations like hemangiomas. Furthermore, IFS may present as a substantial tumor-related bleeding and resemble a kaposiformhemangioendothelioma, which is Kasabach-Merrit syndrome (a fast-growing vascular tumor, thrombocytopenia, microangiopathic hemolyticanemia, and consumptive coagulopathy) [8,10]. IFS mimics other tumors frequently, which is thought to be one of the reasons it is so uncommon. This could potentially make a proper diagnosis more difficult.

Several abnormalities specific to congenital infantile fibrosarcoma may be found using the cytogenetic analysis of fluorescence in situ hybridization (FISH) and real-time polymerase chain reaction (RT-PCR). IFS showed evidence of a distinct reciprocal translocation, t(12;15)(p13;q25), which led to the fusion of the ETV6 and NTRK3 genes. The ETV6/NTRK3 protein interacts with numerous signaling cascades, such as PI3KAKT and Ras-MAP kinase, through the adaptor IRS-1. It may affect a number of cell lineages, such as breast epithelial cells, hematopoietic cells, and fibroblasts. This translocation thus turns into a pathognomonic characteristic of infantile fibrosarcoma [8].

It has previously been determined that positive postoperative margins are a highly significant predicting factor. our patient's proximal margins were positive. Children whose malignancies are completely removed have a 100% 5-year survival rate, according to research. On the other hand, a 76% 5-year survival rate was observed for patients whose illness persisted following resection and adjuvant treatment [6,7,11].

A seven to thirty four percent of patients report having a local recurrence. Older people have a far higher likelihood of recurrence than younger people do. Recurrence in this instance happens 20 days after the initial resection.[12]

At the time of diagnosis, distant metastasis was only recorded in 4% of cases, making it an unusual occurrence. Younger than five-year-olds are susceptible to tumor recurrence, but their patients have a less than 10% metastasis rate. At a 5-year follow-up, 50% of children 10 years of age or older had metastatic spread.[7,12] There were no metastases discovered in our patient. The prognosis for infantile fibroracoma is favorable. An 80–100% five-year survival rate has been recorded.[11] Nowadays, surgery with a broad local excision is the primary treatment in the majority of cases, and it occasionally results in amputation. It has been demonstrated that radiation and chemotherapy can help reduce the bulk of tumors, particularly those that are metastatic, recurrent, or unrespectable. Chemotherapy is recommended for older children to reduce the risk of metastases.[6,7,11] Chemotherapy was used in this instance after surgery for two reasons: 1. The child's age was nine. 2. The tumor was swapped out. Complete remission was attained following therapy.

Actinomycin D, cyclophosphamide, and vincristine is an effective chemotherapy treatment. Vincristin-actinomycin-D treatment was shown to be effective in 71% of patients with infantile fibrosarcoma, according to Orbach et al. [11]

CONCLUSION
Although infantile fibrosarcoma is rare and usually presented after birth, but it is necessary to consider it as a differential diagnose of soft tissue mass in infants and even in children. Patients should be followed up for detecting further relapse or metastasis especially in older ages.

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