International Journal of Contemporary Pediatrics
*Rahul C et al. Int J Contemp Pediatr. 2022 Dec;9(12):1208-1211*
http://www.ijpediatrics.com
pISSN 2349-3283 | eISSN 2349-3291

DOI: https://dx.doi.org/10.18203/2349-3291.ijcp20223070

**Case Report**

# Inflammatory myofibroblastic tumour of an unusual site in an infant

**Rahul Chidurala[1], Latha M. Sneha[2]*, Balaji Thiruvengadam Kothandan[2], Ravikumar Arunachalam[3], Shivpriya Jeyabalakrishnan[3], Vasugi Gramani Arumugam[1], Gokulakrishnan Periyakaruppan[4]**

[1]Department of Pediatric Medicine, [2]Department of Pediatric Hematology and Oncology, Sri Ramachandra Institute of Higher Education and Research, Porur, Chennai, Tamil Nadu, India
[3]Department of ENT, Head and Neck Surgery, SRM Medical College, Hospital and Research Centre, Kattankulathur, Chengalpattu, Tamil Nadu, India
[4]Department of Radiodiagnostics, Sri Ramachandra Institute of Higher Education and Research, Porur, Chennai, Tamil Nadu, India

**Received:** 16 October 2022
**Accepted:** 08 November 2022

***Correspondence:**
Dr. Latha M. S.,
E-mail: drmslatha@yahoo.com



## ABSTRACT

Inflammatory myofibroblastic tumors are rare mesenchymal tumors of intermediate malignant potential, usually seen in the first or second decades of life. They have a high local recurrence rate but rarely metastasize. The involvement of paranasal sinuses or the nasal cavity is relatively uncommon, especially in infants. In children, there is a predilection for the abdominal cavity and it is rare in infants. Complete surgical excision is the treatment of choice. We report a case of a 10-month-old girl with an inflammatory myofibroblastic tumor of the left nasal cavity and sinuses who was initially treated with chemotherapy in view of the complicated surgical process and later underwent near-total resection as she was not responding to chemotherapy. Knowledge of such tumors among physicians and ENT surgeons will improve the prognosis, as completely excised tumors have a very low recurrence rate and adjuvant therapy may not be needed.

**Keywords:** Infant, Mesenchymal tumor, Sinus cavity

## INTRODUCTION

Inflammatory myofibroblastic tumours (IMTs) are low grade neoplasm of mesenchymal origin with an additional inflammatory component comprising lymphocytes and plasma cells.[1] The involvement of paranasal sinuses or nasal cavities is relatively uncommon, especially in infants.[2] Here we report a 10-month-old girl with an inflammatory myofibroblastic tumor of the paranasal sinus and nasal cavity.

## CASE REPORT

A ten-month-old girl born out of a non-consanguineous marriage was brought with complaints of progressive protrusion and watering of the left eye for the past 3 months. She was born by normal vaginal delivery with no significant antenatal or postnatal complications. Her developmental milestones were age-appropriate. On clinical examination, there was epiphora with proptosis of the left eye, with the left eye being pushed inferiorly with increased intercanthal distance. There was no restriction of extraocular movements and pupils were bilaterally

equal and reacting to light. The child did not have any dysmorphic features or neurocutaneous markers. There was no lymphadenopathy or hepatosplenomegaly. Preliminary tests, including a complete blood count, renal function tests, liver function tests, uric acid, LDH, serum alpha-fetoprotein, and beta HCG, all came back normal. MRI of the orbit and CT PNS showed a large T2 hypointense lesion of 2.4×1.8×4.4 cm with an epicenter in the left nasal cavity and ethmoid sinuses. Superiorly, it was seen to cause severe thinning of the roof of the left frontal and ethmoid sinuses; inferiorly, bowing of the hard palate on the left side; medially, deviation of the nasal septum towards the right side; and laterally, protrusion into the extraconal spaces of the left orbit with significant post-contrast enhancement (Figure 1).

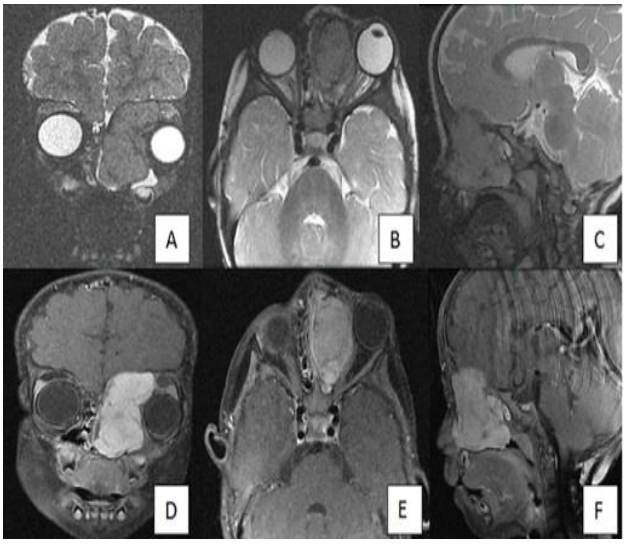


**Figure 1 (A-F): MRI of orbits and paranasal sinuses reveals a large T2 hypointense lesion with epicenter in the left nasal cavity and ethmoid sinuses. Superiorly, it is seen to cause severe thinning of roof of the left frontal and ethmoid sinuses; inferiorly, bowing of the hard palate on left side; medially, devaition of nasal septum towards right side and laterally, protrusion into the extraconal spaces of the left orbit is seen. Significant enchancement of the lesion is seen in post contrast images.**

She underwent a nasal endoscopic biopsy. Histology showed spindle cell proliferation arranged as a storiform pattern and fascicles with mixed inflammatory infiltrates predominantly composed of lymphocytes, plasma cells, neutrophils, and eosinophils (Figure 2 and 3). By immunohistochemistry, the spindle cells were positive for vimentin and focally positive for smooth muscle actin, S-100, and ALK (Anaplastic lymphoma Kinase). (Figure 4) The cells were negative for CD 34 and EMA (Epithelial Membrane Antigen). The Ki-67 labeling index was 5% (Figure 5). With histology and immunohistochemical staining, a diagnosis of inflammatory myofibroblastic tumor was confirmed. Given the risks of surgery and the family's unwillingness to undergo surgery, the child was started on weekly methotrexate and vinblastine chemotherapy following a multidisciplinary tumor board discussion. The response was inadequate with no significant reduction in the size of the lesion after 30 weeks of chemotherapy. Plain MRI of the orbit and PNS revealed a large iso-intense destructive lesion expanding the left ethmoid sinus with large extension in the fronto-ethmoid, inferiorly into the nasal cavity up to the posterior choana crossing the midline with frontal epidural extension. The left frontal sinus had a superior expansion. The family was explained about the possible complications of surgery including damage to lamina payracea, CSF leak, damage to posterior wall of frontal sinus and nasolacrimal duct. During the surgery, the tumor was found occupying the whole left nasal cavity, creating inferior bowing of the hard palate on the left side. Using a lateral rhinotomy incision and a combined endoscopic technique, all of the sinuses were debulked and the tumor was excised completely. Postoperatively, the proptosis has subsided completely. The child has been under regular follow-up for the past 7 months and there is no evidence of local recurrence or distant metastases.

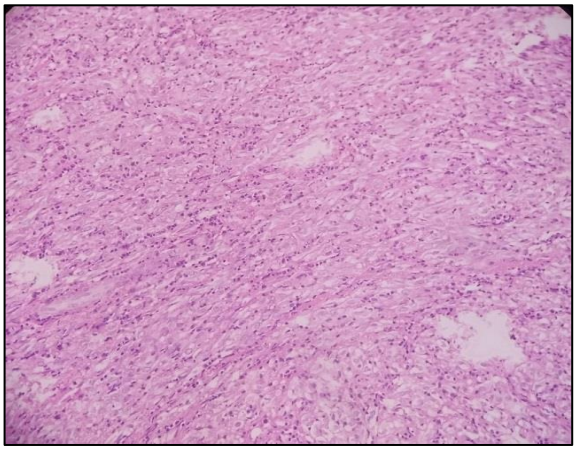

**Figure 2: Spindle cells arranged as bundles and fascicles admixed with inflammatory cells (H and Ex200x).**

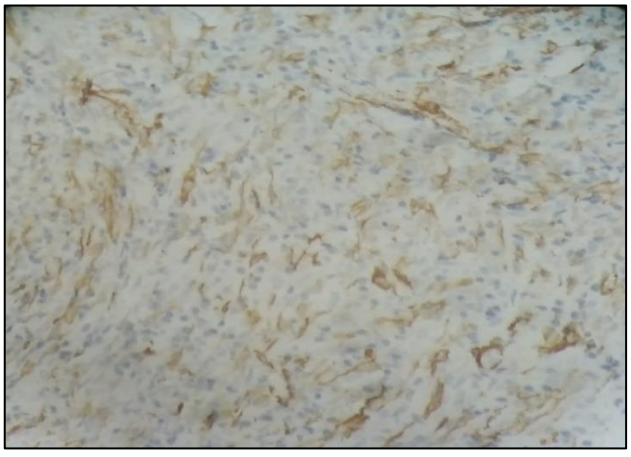

**Figure 3: Spindle cells and inflammatory cells predominantly composed of plasma cells, lymphocytes and occasional eosinophils (H and Ex400x).**

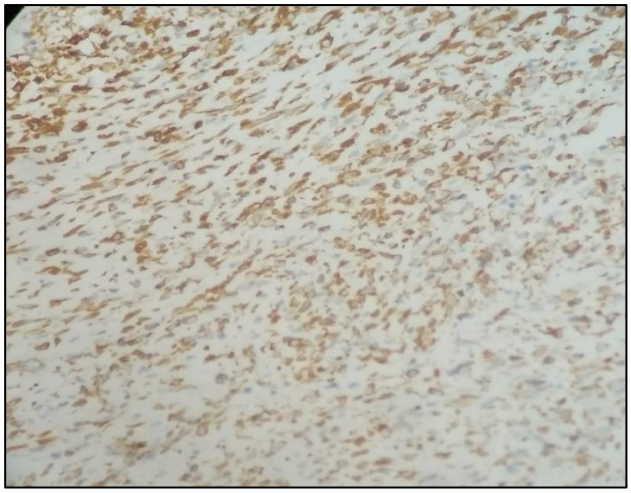

**Figure 4: By IHC, the spindle cells are positive for SMA.**

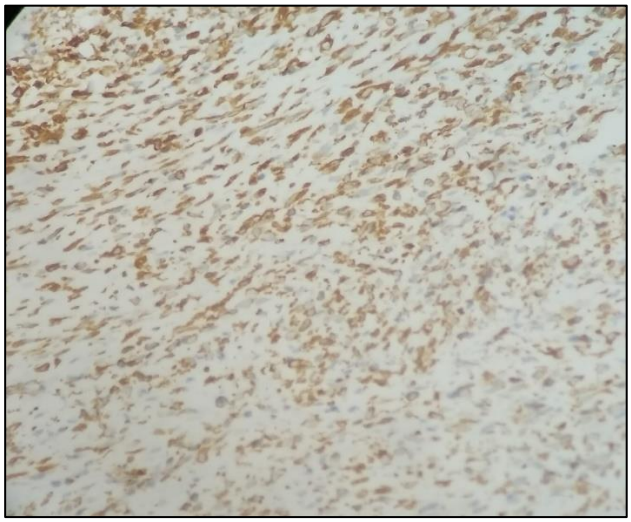

**Figure 5: Spindle cells are positive for ALK.**

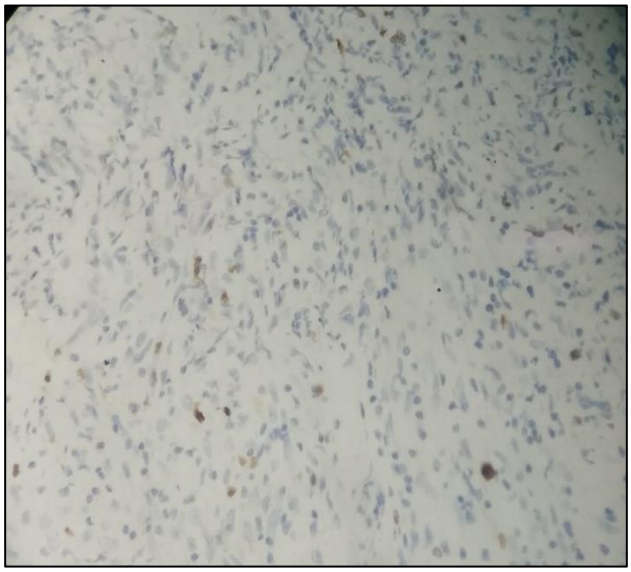

**Figure 6: Ki 67 labelling index - 5% (H and Ex400x).**

## DISCUSSION

The inflammatory myofibroblastic tumor is a mesenchymal tumor with a high local recurrence rate and is a rarely metastasizing intermediate type of fibroblastic or myofibroblastic tumor, according to WHO classification. The aetiology remains largely unclear and has a predilection for children and adolescents.[3] However, its occurrence in infants is very rare. The gastrointestinal tract, lungs, and head and neck area are common areas where IMTs are reported.[2,4] In the head and neck area, the orbit is commonly involved, but occurrences in the nasal cavity or paranasal sinuses are rare.[5] Both benign and malignant tumors occur in the sinonasal area.[6,7] Some of the common benign conditions occurring in the sinonasal area are dermoid cyst, encephalocele, glioma, sinonasal polyp, hemangioma, hamartoma, and leiomyoma.[7,8] The malignant conditions that usually occur in these areas are rhabdomyosarcoma, non-Hodgkins lymphoma, primitive neuroectodermal tumors, nasopharyngeal carcinoma, thyroid carcinoma, and salivary gland carcinoma.[7]

IMTs of the nasal cavity or PNS in infants present quite late until the tumor has grown to block the nasal cavity and has invaded adjusted structures, producing proptosis or disfigurement as in our case. Because of the fibrous nature of the tumor, they are usually hypointense on T2 MRI.[9] Histological differentials include nodular fascitis, solitary fibrous tumor, benign fibrous histiocytoma, myofibroma, and fibrosarcoma.[3] Since IMTs have varied histological presentations, final diagnosis based on exclusion and immune-histo-chemical staining. ALK reactivity found to be associated with local recurrence but not distant metastasis, which associated with ALK-negative lesions, suggesting that ALK reactivity may be favorable prognostic indicator of IMT. Malignant transformation occurs in 8 to 18% of cases, and metastasis has been observed in less than 5% of cases.

Complete surgical removal, whenever feasible, is the treatment of choice. Occurrence of a recurrence depends on the resectability and site of tumor. In a prospective study by the European paediatric soft tissue sarcoma study group (EpSSG), chance of local recurrence is only 12.9% with initial complete excision and a favorable outcome even with near-complete resection.[4] In retrospective study conducted by Kube et al all children who underwent complete surgical resection had no recurrence during follow-up period.[10] In unresectable tumors, a combination of surgery and systemic adjuvant therapy considered. There is no standard recommended adjuvant therapy at present, and the success of each systemic therapy is variable.[4] In EpSSG study, the overall 5-year event-free survival (EFS) for a median follow-up period of 59 months is 82.9% and the overall survival (OS) is 98.1%. In the Kube et al study, overall 5-year EFS is 74% for a median follow-up period of 3.4 years and overall survival is 91%.[10] Hence, overall outcome of IMTs is favorable.

There are only a few case series and case reports on IMTs involving paranasal sinuses. Chun Yan et al retrospectively analyzed 25 cases of IMTs of the nasal cavity and paranasal sinuses.[11] In this study, only 2 out of 25 were from the pediatric age group. ALK1 positivity was reported only in a 2-year-old child (1 out of 25 patients). In this study, 9 patients (9/30, 30%) had local recurrence. Distant metastases were reported in 6 patients (6/30, 20%) and death in 7 cases (7/30, 23.3 %). The higher rates were attributed to the difficulty in complete tumor excision because of the complex anatomical location of the tumor arising from the PNS and nasal cavity. The two pediatric cases were not reported to have local recurrence or distant metastases during the follow-up period.[12] Amaya et al reported a 1 year 11 month child with pediculated IMT arising from the nasal cavity and maxillary sinus. The tumor was completely resected endoscopically and there was no recurrence or metastasis during the follow up period.[5] Cho et al also reported IMT of the nasal cavity in a 4-year-old child which was completely excised and had not received any adjuvant therapy. There was no recurrence or metastasis reported during the follow-up period.[12] In ALK-positive IMTs, targeted therapy with tyrosine kinase inhibitors has shown an adequate response and other novel therapeutic agents are being increasingly tried.[13] The role of radiotherapy in the management of pediatric IMT is not clear and has been largely avoided in most recent studies.

In our case, a radical surgical procedure at 10 months of age was avoided to prevent facial disfigurement in a growing child and underwent surgery at a later stage, to establish a normal nasal airway and prevent functional loss of olfaction and further extension into para nasal sinus.

## CONCLUSION

Although inflammatory myofibroblastic tumors occurring in the sinonasal region are rare, extensive histopathological and immunohistochemical evaluation is essential. Knowledge of such tumors among physicians and ENT surgeons will improve the prognosis, as completely excised tumors have a very low recurrence rate and adjuvant therapy may not be needed.

*Funding: No funding sources*
*Conflict of interest: None declared*
*Ethical approval: Not required*

## REFERENCES

1. Allen CE, Kamdar KY, Bollard CM. Malignant Non-Hodgkin Lymphomas in Children. In: Pizzo PA, Poplack DG, eds. Principles and Practice of Pediatric Oncology. 7th ed. Philadelphia Pa: Lippincott Williams and Wilkins. 2016.
2. Lopez-Nunez O, John I, Panasiti RN. Infantile inflammatory myofibroblastic tumors: clinicopathological and molecular characterization of 12 cases. Mod Pathol. 2020;33:57690.
3. Childhood Soft Tissue Sarcoma Treatment (PDQ®)–Health Professional Version. Available at: https://www.cancer.gov/types/soft-tissue-sarcoma/hp/child-soft-tissue-treatment-pdq. Accessed on 25 October, 2022.
4. Casanova M, Brennan B, Alaggio R. Inflammatory myofibroblastic tumor: The experience of the European pediatric Soft Tissue Sarcoma Study Group (EpSSG). Eur J Cancer. 2020;127:123-9.
5. Alzate Amaya F, Invencio da Costa LF, Martinez Moran A. Unusual Nasal Insufficiency in an Infant: What's Behind the Nostril? Ann Otol Rhinol Laryngol. 2019;128:1078-80.
6. Holsinger FC, Hafemeister AC, Hicks MJ. Differential diagnosis of pediatric tumors of the nasal cavity and paranasal sinuses: a 45-year multi-institutional review. Ear Nose Throat J. 2010;89:534-40.
7. Lazim, NM, Abdullah, B. Multidisciplinary approach to children with sinonasal tumors: A review. Pediatr Invest. 2019;3:173-9.
8. Van Wyhe RD, Chamata ES, Hollier LH. Midline Craniofacial Masses in Children. Semin Plast Surg. 2016;30:176-80.
9. Lai LM, McCarville MB, Kirby P. Shedding light on inflammatory pseudotumor in children: spotlight on inflammatory myofibroblastic tumor. Pediatr Radiol. 2015;45:1738-52.
10. Kube S, Vokuhl C, Dantonello T. Inflammatory myofibroblastic tumors-A retrospective analysis of the Cooperative Weichteilsarkom Studien gruppe. Pediatr Blood Cancer. 2018;65:e27012.
11. Chun-Yan H, Dong GH, Yang DM. Inflammatory myofibroblastic tumors of the nasal cavity and paranasal sinus: a clinicopathologic study of 25 cases and review of the literature. Eur Arch Otorhinolaryngol. 2015;272:789-97.
12. Cho Sung-Il, Choi J Y, Do NY. An inflammatory myofibroblastic tumor of the nasal dorsum. J of Pediatr Surg. 2008;43;e35-7.
13. Mahajan P, Casanova M, Ferrari A. Inflammatory myofibroblastic tumor: molecular landscape, targeted therapeutics, and remaining challenges. Curr Probl Cancer. 2021;45:100768.

**Cite this article as:** Chidurala R, Sneha LM, Kothandan BT, Arunachalam R, Jeyabalakrishnan S, Arumugam VG, et al. Inflammatory myofibroblastic tumour of an unusual site in an infant. Int J Contemp Pediatr 2022;9:1208-11.