Case report: A rare Juvenile Aggressive fibromatosis of the paranasal sinuses

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A case report study

Presentation and history
-1-year-old referred by the neurosurgeons with right nasal mass, rhinorrhoea, epistaxis and right eye proptosis with initial biopsy results of juvenile aggressive fibromatosis . He has a positive family history of aggressive fibromatosis. His aunt was done a lateral rhinotomy and medial maxillectomy for aggressive fibromatosis involving the maxillary sinus.

Examination
-He had a right nasal mass, right eye proptosis and a negative Furstenberg test.

Special investigation
-Ct scan revealed a right ethmoidal sinus mass extending to the right orbit, maxillary sinus, and a defective cribriform plate with intracranial extension.
-There was no intra-axial infiltration. Initial biopsy done by the neurosurgeons came back as juvenile aggressive fibromatosis. An assessment of Juvenile aggressive fibromatosis was made.

Management
-Intra-operative Intranasal endoscopic debulking of the tumour done.
-Repeat biopsy confirmed Juvenile aggressive fibromatosis.
-Post-operative CT scan showed right nasal cavity enlargement and nasal septum deviation to the left. No intra-axial space-occupying lesion seen.
-Patient now receiving adjuvant chemotherapy to complete a course of 43 weeks.

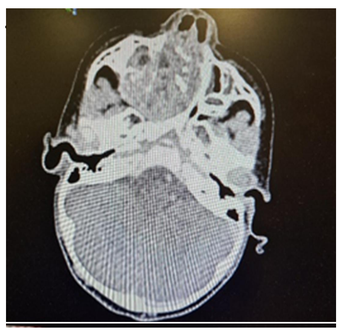
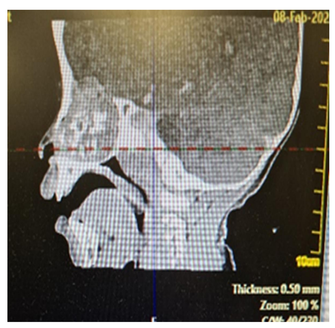


Fig 1. Pre-op CT scan of the paranasal sinuses (Axial and Sagittal view)



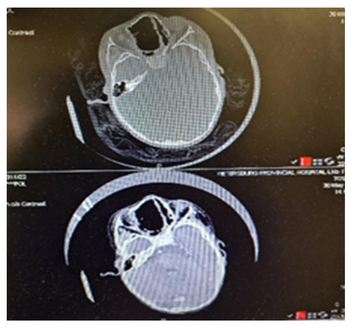


Fig 4. Post-op CT Scan of the paranasal sinuses and the brain (axial view)

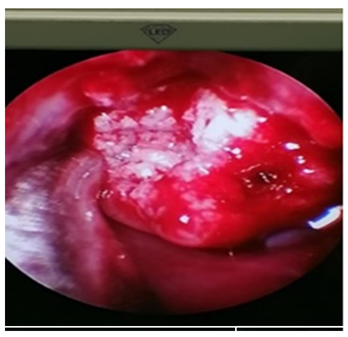


Fig 2. Pre-op findings.

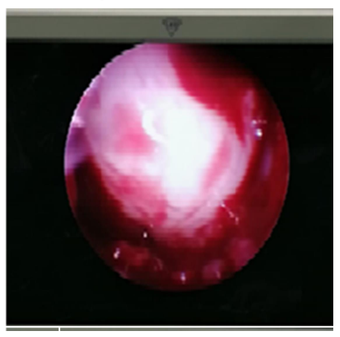


Fig 3. Post-op findings.

Discussion

Juvenile aggressive fibromatosis is a fibroblastic neoplasm, arising from musculoaponeurotic stromal elements [3]. It is characterized by its tendency to destroy adjacent structures and has a high rate of local recurrence [1]. Also called desmoid tumour or desmoid type fibromatosis tumours [3].

Epidemiology
Accounting for approximately 0.03% of all neoplasms, and less than 3% of all soft tissue tumours [1]. Has no significant racial or ethnic predilection. It is seen in the general population at a rate of 2-4 in a million per year. Affects female and male equally at a ratio of 1:1 [2]. It is predominantly seen in paediatrics patient at the age between birth and 16 years, with the mean age being 5 months and 4 years [4].

Pathogenesis
Aetiology of desmoid tumours is not clear but trauma, genetic and endocrine factors have been implicated [3].

Natural history
Are benign, slow growing, bulky tumours that is locally invasive and infiltrates adjacent tissue structure.

Classification
Intrabdominal and extra-abdominal aggressive fibromatosis. The extra-abdominal may occur in the head and neck but is extremely rare in the paranasal sinuses [3].

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Histology
Its characteristics are small bundles of spindle cells in an abundant fibrous stroma [3].

Special investigations:
Computed tomography scan and Magnetic resonance imaging are the best modalities used to identify the primary, residual, recurrent and surveillance of disease[1].

Management
Complete surgical excision with wide margins is the treatment of choice. Primary radiation therapy can be used for unresectable tumour, residual disease and salvage therapy. Adjuvant therapy involves local radiation, NSAIDs and/or systemic cytotoxic and non-cytotoxic agents and is recommended in irresectable tumours and where there is residual tissue after surgical excision[1].

Conclusion

Juvenile aggressive fibromatosis are rare in children and very few cases are seen in the paranasal sinuses. Surgical resection with wide margins remains the primary treatment for extra-abdominal fibromatoses.

Reported cases of pediatric desmoid fibromatosis of the paranasal sinuses and management [1]

Age	Location	Presentation	Pathology	Therapy	Response
2-year-old male	Right maxillary sinus	Nasal obstruction	Aggressive fibromatosis	Surgical resection	Lost to follow up
14-year-old female	Right parotid/ mandible Right	facial deformity	Aggressive fibromatosis	Surgical resection (positive margins)	(No recurrence at < 1 year
15-month-old male	Nasal cavity/ anterior maxilla	Facial deformity	Aggressive fibromatosis	Surgical resection (positive margins) 2. Surgical resection (negative margins)	. Recurrence in 1 month, no recurrence
2-year-old male	Left maxillary sinus	Nasal deformity	Desmoid fibromatosis	Surgical resection (twice), followed by adjuvant tamoxifen	No recurrence at 2 years

Conley et al. [2] reported a series of 40 different cases, three cases between the ages 1–10. One of these cases involved the ethmoid sinus. Fu [3] reported two cases of juvenile fibromatosis ages 2 and 10. One of these cases involved the maxillary

References:

1.Shaheen E Lakhan et al. **Aggressive juvenile fibromatosis of the paranasal sinuses: case report and brief review**. Hematol Oncol. 2008
2. lharle@gnif.org https://www.cancer.gov/pediatric-adult-rare-tumor/rare-tumors/rare-soft-tissue-tumors/desmoid-tumor.
3. Astrid L. Kruse et al. **Aggressive fibromatosis of the head and neck: a new classification based on a literature review over 40 years (1968–2008)**. Oral and Maxillofacial Surgery,14(4):227-232. 2010
4. Keerthi Krishnankutty Nair et al. **Aggressive fibromatosis of the oral cavity in a 5-year-old boy: a rare case report**. Pan Afr Med J. 2017