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RESEARCH ARTICLE

LOW-GRADE SINONASAL FIBROSARCOMA : A CASE REPORT

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Abstract

Fibrosarcoma is a rare malignant tumor. The sinonasal localization is extremely uncommon.Theclinical presentation is similar to other neoplasms of this region, consisting of nonspecific symptoms such as epistaxis and nasal obstruction. Definitive diagnosis is obtained on the basis of histopathology and immunochemistry.This isthe case of a 40-year-old woman with a six months complaint of left nasal obstruction, recurrent epistaxis, hyposmia and cephalalgia. Nasal endoscopy revealed a regular pink soft-tissue mass filling the left nasal cavity. CT scan and MRI showed a well-defined, irregularly contoured tissue tumor,occupying the left nasal fossa, with matrix of calcifications within the tumor.A biopsy was performed, and the histopathological findings revealed a low-grade fibrosarcoma with spindle-shaped cells arranged in a 'herringbone' pattern. The immunohistochemistry was positive for vimentin staining. The tumor was completely resected with free margins using the endoscopic endonasal approach. No adjuvant radiotherapy was prescribed.The patient is disease-free at a regular five-year postoperative follow-up.Only few cases of sinonasal fibrosarcoma has been reported in literature. There are no specific symptoms or imaging features. Still, this is the first case describing intra-tumoral calcifications. The mainstay of treatment is surgical excision with radical margins.Radiotherapy may be adjunctive in cases of positive margins or incomplete resection.

Introduction:-

Fibrosarcomas are rare malignant tumors of mesenchymal origin that preferentially develop in the extremities(1).Approximately 2% to 15% of all sarcomas arise in the head and neck region, representing about 1% of all malignancies of this area(2). Only 7 to 10% of these sarcomas are fibrosarcomas(1). The sinonasal localization is even rarer. There are few cases that have been reported in literature. The clinical presentation of SinonasalFibrosaroma (SNFS) is similar to other neoplasms occurring in this region, consisting on nonspecific symptoms such as unilateral nasal obstruction and epistaxis(3). After nasal endoscopy and imaging, the definite diagnosis is based on histopathological features and immunohistochemistry (4). Due to the high risk of local recurrence and the low risk of distant metastasis, the mainstay of the treatment of SNFS is wide local excision with large tumor-free margins(1,3). We report a patient with SNFS, which was completely excised by endoscopic endonasal surgery.

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Case Report:
A 40-year-old female patient presented to our hospital with six months complaint of left nasal obstruction, recurrent epistaxis, hyposmia andcephalalgia. She had no medical history. She also had no history of radiation therapy or any surgical procedure. Physical examination did not show any abnormalities. There were no ocular or neurological sign, as well as no palpable lymph nodes in the neck. Nasal endoscopy revealed a regular pinksoft-tissue mass filling the left nasal cavity and pushing back and reducing the middle turbinate. The mass appeared to originate from the olfactory cleft (Figure 1). The nasopharynx, oral cavity and pharyngo-larynx were free of other pathological findings. Routine hematological and biochemical tests were normal.

Craniofacial computed tomography (CT) scan revealed a well-defined, irregularly contoured tissuetumor, measuring 43 x 35 x 38 mm, centered in the left nasal fossa and extending into the left ethmoid cells. There were matrix calcifications within the mass. This tumor thins and displaces the medial wall of the maxillary sinus and the homolateral orbital plate, lyses the nasal septum and the perpendicular ethmoidal lamina, and extends discreetly into the right superior and middle turbinates (Figure 2A). Axial and coronal craniofacial MRIsections show the tumor exhibiting a discrete heterogeneous iso-signal intensity in T1-weightedimages, with marked enhancement after gadolinium injection, andheterogeneous intermediate low-signal intensityT2-weightedimages, with no extension to thehomolateral maxillary sinus or orbit (Fig. 2B,C,D).There were no orbital or intracranial extension.No lymphadenopathy wasfounded at the level of the cervical segments.

A biopsy was performed under local anesthesia, and the histological examination revealed a low-grade fibrosarcoma with spindle-shaped cells arranged in a 'herringbone' pattern. These cells showed low mitotic activity (< 1 mitoses per 10 high-power fields) and were frankly positive for anti-vimentin antibodies (Figure 3). The tumor extension assessment found no distant metastases.

The tumor was completely resected with free margins using the endoscopic endonasal approach. No reconstruction was necessary. The immediate postoperative course of the patient was uneventful. The patient was discharged home on postoperative day 5.

Our patientdid not receive adjuvant radiotherapy on the decision of themultidisciplinary consultation meetingwhich included surgeons, oncologists,radiotherapists and pathologists. A regular follow-uphas beenscheduled. Five years after surgical removal of the tumor,her post-operative endoscopic examinations and CT scans have shown no evidence of tumor recurrence.

Discussion:-

Fibrosarcoma is a malignant tumor that develops from fibroblasts(5). Its sinonasal localization is uncommon, as only 16% of fibrosarcomas arise in the head and neck region, with the main predilection site being the extremities(6). There are few cases reported in the literature. In a systematic review, Bughara et al. count a total of 109 cases that have been reported in the English language literature between 1952 and 2019 (7). The largest serieswas a retrospective cohort including 51 patients (8).

In early reports, SNFS usually presented in the third and fourth decades of life, more commonly among males(1). However, in Bughara's systematic review, which included the largest series, it was shown that SNFS often presents in the fifth to seventh decades of life, with both sexes equally represented(7).The majority of cases are sporadic and the etiology remains uncertain. Radiation exposure have been reported to be the most important etiological factor for SNFS, followed by underlying bone diseases like chronic osteomyelitis, Paget’s disease, fibrous dysplasia(5,9). Although, SNFS can arise from anywhere in the nasal cavity and paranasal sinuses, the most frequently reported site of origin is the maxillary sinus (8). In Patel’s cohort, SNFS is located in the maxillary sinus in 54,9% of cases, followed by the nasal cavity in 23.5% of cases (8).

The most common presenting symptoms are nasal obstruction, epistaxis, hypoesthesia and facial pain(7). These nonspecific symptoms are present in a wide range of benign and malignant diseases of the nasal cavity and paranasal sinuses. Suspicion of malignancy should be raised by the unilateral pattern and lack of improvement with treatment; hence, further investigations must be carried out.SNFS’s endoscopic aspect varies in literature, it could be a polypoidal smooth soft-tissue mass(10–12) or a dark blue hemorrhagic mass (5).Both CT with contrast agent and MR imaging are usually required to fully evaluate the bone and soft tissue characteristics of the lesion. The paucity of SNFS cases within the literature makes studies examining their imaging features rarer. Zheng et al. published a

study, in which they described the common CT and MRI characteristics found in 7 patients with confirmed SNFS (4). These tumors present usually as a single lobulated or irregular heterogeneous soft-tissue density mass with areas of patchy fluid attenuation.The margins can be either well- or ill-defined on CT and MRI images. In addition, suspicion of SNFS should be increased when the lesions showed characteristic signal change on MRI. On T1 weighted imaging, the tumors commonly tended to be isointense or mildly hypointense, and showed heterogeneous mild hypointensity on T2 weighted imaging (4). Expansive or osteolytic bone destruction and heterogeneous delayed contrast enhancement may be observed (4).

The definite diagnosis is only obtained on pathological examination. Thesetumors are typically unencapsulated, densely proliferating and slow-growing spindle cell neoplasms(13), that appear to originate in the periosteum rather than in mucosal connective tissue(12). SNFS shows variable collagen production and has the ability to metastasize. The neoplasm does not show differentiation in other types of tissue (14). Microscopically, SNFS is hypercellular neoplasm composed of thin, elongated spindle cellsarranged in fascicles and bundles intersecting at various angles, called the herringbone pattern, associated with nuclear atypia and large number of mitoses(3,14). Areas of prominent necrosis might be seen. Immunohistochemistry staining is positive for Ki67, P53, and vimentin. SNFS is negative for pancytokeratin, desmin, S100 and SMA. These histopathological features are helpful to distinguish SNFS from other differential diagnoses such as benign sinonasal polyps, fibroma, inflammatory myofibroblastic tumor, Ewing's sarcoma, lymphoma, sinus cancer (adenocarcinoma and squamous cell carcinoma), and desmoid fibrosarcoma. It is important to mention that the majority of SNFS cases are low-grades(8). Despite the rarity of distant metastasis, local recurrence is frequent (7).

Surgery remains the commonly accepted treatment of choice(3). A wide local excision with an extensive surgical margins is generally recommended (7). Endoscopic approach has been well reported, with good oncologic outcomes and less morbidity than radical open approaches (15). However, the endoscopic approach does not always permit an “en bloc resection” of the tumor with radical margins (15,16).Adjuvant radiotherapy is indicated in cases with positive surgical margins or macroscopically incomplete excision (1). Chemotherapy has been performed in combination with radiotherapy as palliative therapy for inoperable patients (5).For unresectable tumors, it is suggested that neoadjuvant chemotherapy should be used preoperatively to reduce tumor size (7).

The prognosis of SNFS depends on the degree histological differentiation grade, the tumor size, and the surgical treatment outcome, especially the statute of the surgical margins (17). The 5-year survival rate is low, ranging from 20% to 35% (5,7). Death often results from intracranial extension of a persistent or recurrent tumor (16). Due to its high rate of local recurrence, frequent and regular endoscopic and imaging controls are required for early detection of recurrences(16).

Conflict of Interest :
None.

Conclusion:-

In this report, we present a case of a low-grade sinonasal fibrosarcoma (SNFS) thatwas completely resected endoscopically without complications or recurrence after five-year follow-up. This rare sinonasal malignant neoplasm is diagnosed by its histopathological features andimmunohistochemistry. The unusual feature of SNFS in our case was the presence of matrix calcification within the tumor as seen on the CT scan.To our knowledge, this feature has never been mentioned in the few cases that have been reported in the literature.

Figures:

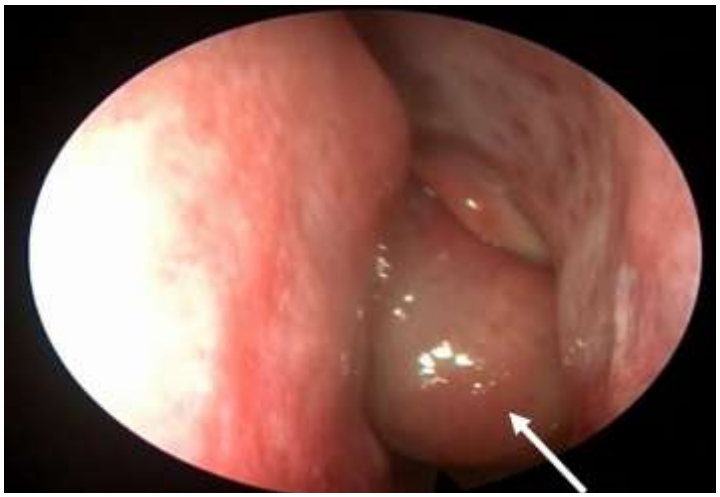

Figure 1:- Endonasal endoscopic view showing the soft-tissue tumor.

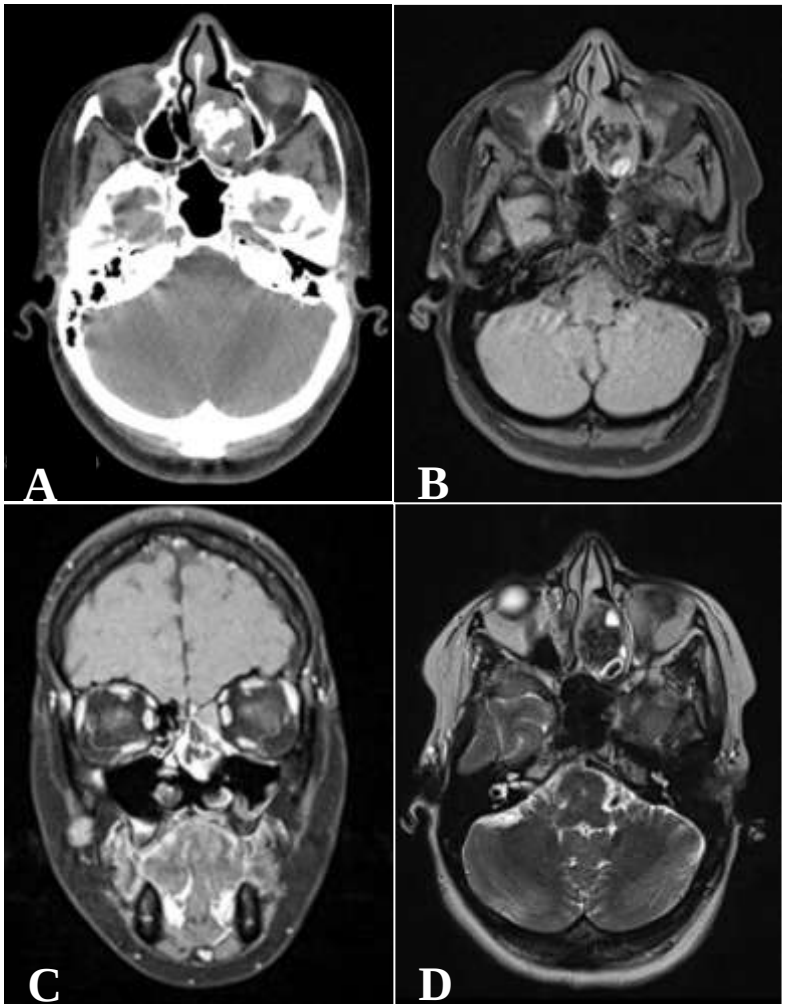

Figure 2:- (A) Axial CT scan showing the well-defined, irregularly contoured tissue tumor with in the left nasal cavity. (B,C) Axial and coronal MRI T1-weighted after gadolinium injection showing that the tumor occupies the left nasal cavity and exhibits a discrete heterogeneous iso-signal, with marked enhancement after gadolinium injection. (D) Axial MRI T2-weighted showing an heterogeneous intermediate low-signal intensity tumor.

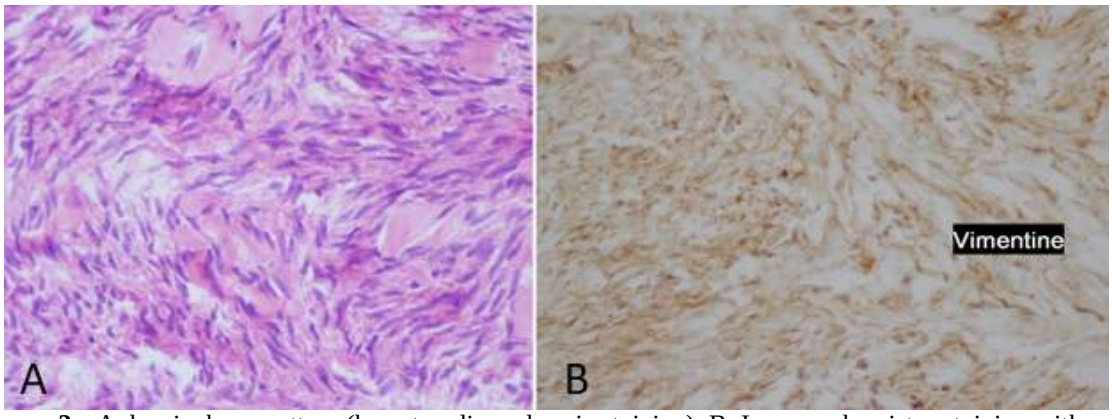

Figure 3:- A: herringbone pattern (hematoxylin and eosin staining), B: Immunochemistry staining with anti-Vimentin antibodies(V9-BiosB) frankly positive.

References:-

1. Koka V, Vericel R, Lartigau E, Lusinchi A, Schwaab G: Sarcomas of nasal cavity and paranasal sinuses: chondrosarcoma, osteosarcoma and fibrosarcoma. J Laryngol Otol. 1994, 108:947-53. 10.1017/s0022215100128609.
2. O'Neill JP, Bilsky MH, Kraus D. Head and Neck Sarcomas. Neurosurg Clin N Am. janv 2013;24(1):67-78.
3. Perez-Ordonez B, Huvos AG. Nonsquamous lesions of nasal cavity, paranasal sinuses, and nasopharynx. In: Gnepp DR (ed). Diagnostic Surgical Pathology of the Head and Neck. Philadelphia: W.B. Saunders Company; 2001:79–89.
4. Zeng J, Liu H, Liu L, Liao W, Hu P, Wang X, et al. Fibrosarcoma arising in the paranasal sinus: a clinicopathological and radiological analysis. DentomaxillofacialRadiol. juill 2018;47(6):20170459.
5. Maliki O, Aleksandrov O, Carles P, Planquart X, Vaz E, Bertrand C, et al. Fibrosarcoma of the nasal cavity: A case report. Egypt J Ear Nose Throat Allied Sci. nov 2014;15(3):275-7.
6. Batsakis JG, Rice DH, Howard DR. The pathology of head and neck tumors: spindle cell lesions (sarcomatoid carcinomas, nodular fasciitis, and fibrosarcoma) of the aerodigestive tracts, Part 14. Head Neck Surg. 1982;4(6):499–513.
7. Bughrara MS, Almsaddi T, John J, Prentice B, Johnson J, Henriquez O, et al. Fibrosarcomas of the Paranasal Sinuses: A Systematic Review. Cureus. 10 août 2022;14(8).
8. Patel TD, Carniol ET, Vázquez A, Baredes S, Liu JK, Eloy JA. Sinonasal fibrosarcoma: analysis of the Surveillance, Epidemiology, and End Results database. Int Forum Allergy Rhinol. févr 2016;6(2):201-5.
9. Zhang P, Zhao L, Zhu YJ, Qiu B, Guo SP, Li Y, et al. Prognosis of Fibrosarcoma in Patients With and Without a History of Radiation for Nasopharyngeal Carcinoma. Ann Surg Oncol. févr 2017;24(2):434-40.
10. Agarwal MK, Gupta S, Gupta OP, Samant HC. Fibrosarcoma of nose and paranasal sinuses. J Surg Oncol. sept 1980;15(1):53-7.
11. Zouhair N, Chaouki A, Ballage A, Abada RE, Rouadi S, Roubal M, et al. Fibrosarcoma of the ethmoid sinus: A rare entity. Int J Surg Case Rep. 2019;59:136-9.
12. Alameda YA, Perez-Mitchell C, Busquets JM. Nasal Cavity Ossifying Fibrosarcoma: An Unusual Fibro-Osseous Neoplasm. Ear Nose Throat J. nov 2010;89(11):E1-3.
13. Heffner DK, Gnepp DR. Sinonasalfibrosarcomas, malignant schwannomas, and ''Triton'' tumors. A clinicopathologic study of 67 cases. Cancer. 1992;70(5):1089–1101.
14. Fu YS, Perzin KH. Nonepithelial tumors of the nasal cavity, paranasal sinuses, and nasopharynx. A clinicopathologic study. VI. Fibrous tissue tumors (fibroma, fibromatosis, fibrosarcoma). Cancer. 1976 Jun;37(6):2912-28. doi: 10.1002/1097-0142(197606)37:6<2912::aid-cncr2820370649>3.0.co;2-l.
15. Bercin S, Muderris T, Kiris M, Kanmaz A, Kandemir O. A Rare Sinonasal Neoplasm: Fibrosarcoma. Ear Nose Throat J. mai 2011;90(5):E6-8.
16. Plaza G, Ferrando J, Pinedo F. Sinonasal fibrosarcoma: a case report. Eur Arch Otorhinolaryngol. juill 2006;263(7):641-3.
17. Wadhwan V, Chaudhary MS, Gawande M. Fibrosarcoma of the oral cavity. Indian J Dent Res. 2010;21:295–298.