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Fibrosarcoma of Nose and Paranasal Sinuses

(Case Report and Review of Literature)

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SUMMARY

Fibrosarcoma of nose and paranasal sinuses is uncommon and a few cases have been reported to date. Because the histological diagnosis is difficult, these tumors may be misdiagnosed and confused with other tumors. Early signs and symptoms of the tumor are vague, delayed and at times misleading. We report a case of fibrosarcoma of nose and paranasal sinuses in a 50 year old male with a brief review of literature.

CASE REPORT

The patient was a 50 years old male who came to the department of E.N.T with complaints of swelling in the palate of 3 months duration, swelling in the submandibular region for the past one year, right sided nasal blockage, epistaxis and purulent rhinorrhoea for the past 4 years. The patient was a smoker for the past 10 years and worked in a copper wire manufacturing factory.

He had a history of previous surgery on his nose which was done 4 years back and which relieved his symptoms at that time. The nature of the surgery was not available. He was biopsied two years later due to recurrence of nasal blockage and epistaxis and a Histological diagnosis of Soft Tissue Sarcoma of right nostril was made. He was given radiotherapy but his symptoms did not improve. He was then advised surgery, which he ignored and instead started with homeopathic treatment. Interestingly the epistaxis stopped and never recurred. During this time he developed an enlarged submandibular lymph node. For this he was given chemotherapy for 4 months and the lymph node disappeared but the nasal symptoms continued. During this time he noticed a swelling eroding the palate and the patient lost 3 of his right upper molar teeth.

On physical examination, he was found to have a huge swelling blocking the right nostril. The swelling was firm in consistency and looked fleshy. It had pushed the septum to the left side. The patient also had a 3 cm diameter, firm fleshy swelling involving the hard palate and posterior part of right upper alveolus.

CT scan of paranasal sinuses showed a large soft tissue homogeneously enhancing mass in the right maxillary antrum, destroying its medial wall and extending into the nasal cavity. Inferiorly, it involved part of hard palate with soft tissue extension into the right cheek. Superiorly, it was extending upto the posterior ethmoid cells and there was doubtful involvement of superior orbital fissure.

Right total maxillectomy was performed using Weber Ferguson incision. A huge mass was found extending into the nasal cavity and filling the maxillary antrum. The mass had eroded the hard palate and the posterior part of right upper alveolus. The whole right maxilla was removed.

The microscopic examination of the excised specimen showed a highly cellular tumor composed of pleomorphic spindle shaped cells (Fig 1 and Fig 2), and many bizarre giant cells, some showing multinucleation (Fig 3). The atypical tumor cells at places showed a herring bone and storiform arrangement with focal myxoid change. These appearances were diagnosed as a poorly differentiated fibrosarcoma. Resection margins were clear on histological examination.

The patient was alright 4 months postoperatively when he developed a rapidly enlarging mass at his right temple. There was no recurrence at the site of surgery. He was given radiotherapy to the temple and the results are awaited.

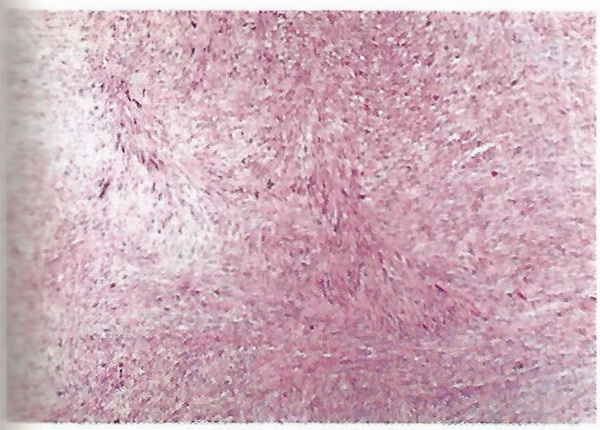

Fig 1: Fibrosarcoma, Low Power View (H&E). Bundles of spindle shaped cells are seen running in various directions. Hyperchromatic nuclei can be seen on this power.

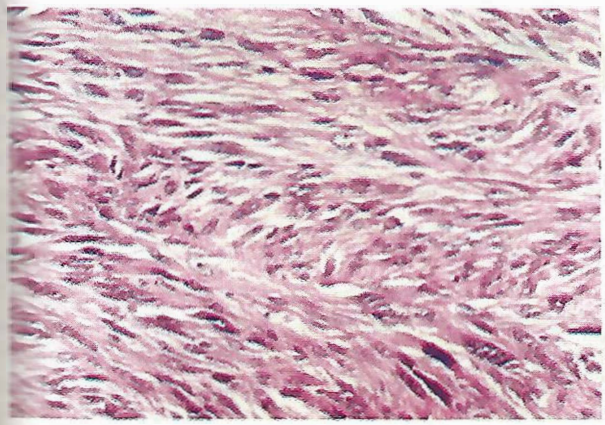

Fig 2: Fibrosarcoma, Medium Power View (H&E). This section shows neoplastic fibroblasts with thin elongated nuclei. Some large nuclei with hyperchromatism are also seen in the lower right field.

DISCUSSION

Fibrosarcoma of the nose and paranasal sinuses is an uncommon malignant tumor.

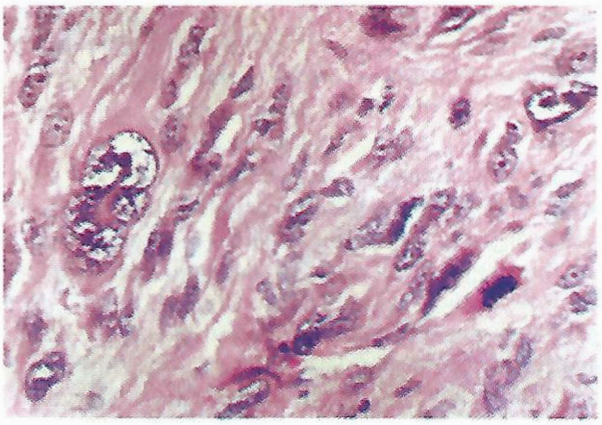

Fig 3: Fibrosarcoma, High Power View (H&E). This section shows pleomorphism and a giant multinucleate tumor cell. A mitotic figure is seen in the right lower corner.

On reviewing the literature, we found that in 1969 Lewis[1] found only 52 cases of sarcoma in 514 cases of malignant tumors of nose and paranasal sinuses.

In 1973, Cronin[2] reported two cases of fibrosarcoma involving the maxillary sinuses and one involving the frontal sinus.

In 1974, Fu and Perzin[3] reported 13 cases of fibrosarcoma in 256 cases involving non-epithelial tumors of the nasal region, sinuses and nasopharynx.

In 1980, Agarwal and colleagues[1] reported two cases having this malignancy. In 1981, Broniatowski and Haria[5] reported two cases of fibrosarcoma of the nose and paranasal sinuses. In 1989, Olekszyk and colleagues[6] reported one case of fibrosarcoma of nose and paranasal sinuses.

No definite cause of fibrosarcoma has been found though previous trauma[7], nasal polyposis and history of radiation therapy have been implicated in the development of fibrosarcoma. No male or female predominance has been demonstrated[6].

Stout divided fibrosarcoma into two histological types based on their degree of differentiation. Well differentiated lesions have few or no mitotic cells, well developed collagen and abundant reticulin fibres. Poorly differentiated fibrosarcomas have marked anaplastic appearance, increased mitosis and

abundant hyperchromatism. There is no correlation between microscopic appearance and prognosis.

Distant spread is mainly haematogenous and the most common site is Lungs. In a study by Van der Werf-Messing and Van Unnik, lymphatic spread occurred only in 12 out of 139 cases (8.6%) and this was more common with extensive lesions[8].

As far as treatment is concerned, wide excision is necessary but surgical mutilation should be avoided[9]. Haematogenous metastasis is a frequent cause of death[10]. In adult fibrosarcoma, at all sites, local recurrence rates are high[11].

There is little data to support the efficacy of chemotherapy, but in our case the lymph node in the neck disappeared following chemotherapy but the nasal symptoms persisted. Radiation therapy has been the only modality other than surgery to increase the survival rate[6]. Windeyer and colleagues report a 5 year survival rate of 50% in 58 cases.

Sears and colleagues[12] have shown that grade and size of tumor are of prognostic significance.

Russel and colleagues[13] have reported that when fibrosarcoma invaded bone, major nerves or blood vessels (Stage 4), the 5 year survival rate dropped to less than 10%. However, in those cases where regional lymph nodes are affected but bones are not, the 5 year survival rate increases to 43%.

Early diagnosis and aggressive surgical treatment or radiotherapy or both are necessary for good prognosis but early signs and symptoms are vague and diagnosis is delayed as in any malignancy of paranasal sinuses[6].

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