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SYNOVIAL SARCOMA OF NASAL CAVITY AND THE ETHMOIDAL SINUS: REPORT OF A RARE LOCALIZATION

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Abstract:-

Introduction: *Synovial sarcoma is a highly malignant tumor affecting mostly extremities of young adult. The nasal location represents less than 1% cases. Only a few sporadic cases have been reported. Its treatment is not well established.*
Case report: *We report the case of a synovial sarcoma of nasal cavity in a 16 years old patient characterized by its recurrent character being controlled by a treatment combining surgery and chemoradiotherapy.*
Discussion: *We will discuss the various radio-clinical, histological and therapeutic particularities referring to a literature review.*

Key words *Synovial sarcoma; sino- nasal; endoscopic endonasal Surgery*

BACKGROUND:-
Synovial sarcoma is a high-grade, soft tissue, malignant disease associated with poor outcome. Typically, it involves the extremities, with less than 10 per cent occurring in the head and neck region [1]. The tumor in the nasal cavity is less than 1%[2]. The clinical and radiological signs are not pathognomoniques. The diagnosis of certainty is histological associated with the immunohistochimie. The treatment of choice for synovial sarcoma of the head and neck is a complete surgical excision of the tumour mass followed by adjuvant radiotherapy. We report a new case of nasal synovialosarcoma where the histological study caused a problem of differential diagnosis with other types of sarcomas.

Clinical Case
M, E aged 16 years, followed for dorso-lumbar scoliosis, has been presenting 2 months ago a left nasal obstruction with epistaxis and bloody rhinorrhea. The examination revealed a tumor of the left nasal fossa exteriorized by the left nostril, reddish, covered with a smooth mucous membrane, bleeding on contact (Fig. 1), of hard consistency in places. Palpation of the nasal pyramid was painful. the patient didn't presents neurological signs nor cervical lymph nodes. Ophthalmologic examination found normal visual acuity and ocular mobility on both sides. CT of the facial mass found a heterogeneous hypodense left nasal process heavily enhanced after injection of contrast product. The tumor infiltrates the ethmoidal cells and fills the nasopharynx on the left side. The patient underwent complete endoscopic removal of the tumor. Anatomopathological examination is in favor of a sarcoma. An angiosarcoma, osteosarcoma and leiomysarcoma are discussed. The immunohistochemical study supports a synovialosarcoma (Figure 2, 3, 4). Molecular biology has not been realized for soscioeconomic reasons.The extension report included cervico-thoraco-abdominal CT showed no metastases. After the delay of the histological diagnosis, the patient begins her radiotherapy. locoregional two months later the patient presents a recurrence: the tumor is extended to the contralateral nasal fossa, to the right maxillary sinus, to the homolateral orbit without infiltration of the ocular muscles (Fig.5) .After a multidisciplinary consultation, the patient was reoperated endonasally (naso-ethmoidal and maxillary left) with simple surgical procedures followed by adjuvant radiotherapy: 33 Gy at 2Gy / session and chemotherapy: 6 courses of Adriamycin And Isofosfamide. The nasal endoscopy control is without anomaly. The control CT finds a laminar material in the residual cavity of the left nasal cavity evoking a parasitic graft (Fig. 3) for which a treatment is prescribed. The evolution is marked by a good locoregional control with a remission of 2 years.

Discussion
The tissue origin of synovialosarcoma is still unknown and its risk factors are not clearly established [3]. The sinonasal localization of this tumor is exceptional. Synovial sarcomas show a male predominance (3:2) and predilection for patients between 25 and 36 years of age [4]. Clinically, the symptoms are those of malignant tumors: nasal obstruction, rhinorrhea and epistaxis. At an advanced stage the tumor syndrome appears with invasion of the noble structures (orbit, skull base and infratemporal area). This was not the case with our patient. Imaging is not specific. The CT and MR imaging appearance of synovial sarcomas of the head and the neck have been described as most often well limited with sometimes a cystic or hemorrhagic component and calcifications. CT classically displays a multilocular tumor with smooth margins and heterogeneous enhancement after injection of contrast medium. MR imaging revealed a tumor of intermediate intensity on T1-weighted sequences and of variable intensity on T2-weighted sequences, with heterogeneous enhancement after injection of contrast material [5]. The assessment of locoregional invasion is imperative by imaging. The diagnosis of malignancy is confirmed by histology with immunohistochemistry. Synovial sarcoma classically demonstrates a biphasic pattern consisting of both spindle cells and epithelioid cells or monophasic consisting solely of epithelial or fusiform cells [6]. Immunohistochemically, synovial sarcomas are immunoreactive for cytokeratins, EMA, S100, bcl-2, CD99, calponin [7]. Genetics have a fundamental contribution since synovialosarcoma is specifically associated with a chromosomal translocation t (X; 18) (p11.2; q11.2) whatever its location [5] [6].it was not available for our patient. The differential diagnosis is mainly with rhabdomyosarcoma and fibrosarcoma. However, it is not easy to distinguish it from LMNH and other types of sarcomas such as liposarcoma and leiomyosarcoma[5]. Due to the aggressive nature of the tumor, complete surgical resection (adapted to the tumor site and its extension) remains a key condition to prevent locoregional recurrences and metastases. The surgical approach was not evaluated in the cases reported. There is no prophylactic lymph node dissection in the absence of cervical adenopathies[9]. Several series have reported the benefit of postoperative radiotherapy that is correlated with longer survival and a lower rate of recurrence [10]. However, there is no study showing a statistically significant difference showing the superiority of radiosurgical association compared to surgery alone [8]. In addition to surgery and radiotherapy, chemotherapy has been studied to improve outcomes, but its role remains controversial. Some studies report good responses to chemotherapy, others have refuted these results. However, these studies have focused on a small sample of patients [9]. Our patient has evolved well after complete removal of the tumor followed by radiochemotherapy with a current 2 years follow up. The reported 5-year survival rates for patients with synovial sarcomas range from 36 to 76%; the 10-year survival rates range from 20 to 63%*[7]*. Patients with a tumor size greater than 5 cm have a higher risk of local recurrence, metastasis and mortality [10]. Other prognostic factors are incriminated as the age of the patient, the number of mitoses, the presence of necrosis, the monophasic or biphasic histological type [10] and the degree of tumor extension [4].

Conclusion
The nasal location of synovialosarcoma is rare. It has no radio-clinical specificity. It is necessary to consider it in front of the signs of malignancy of a nasal tumor. Diagnosis must be early and certified by genetic study. Given the small

number of cases reported, its treatment is poorly coded based on surgery and postoperative radiotherapy. Chemotherapy remains a therapeutic option being evaluated.

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Figure 1: Left nasal tumor haemorrhagic outgrowth by the nostril

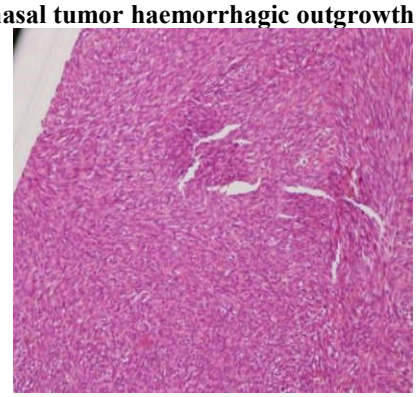

Figure 2: Monomorphic proliferation of spindle shaped cells arranged in fascicles with hyperchromatic nuclei, rarely prominent nucleoli and small amounts of cytoplasm many dilated thin wall blood vessels in the stroma

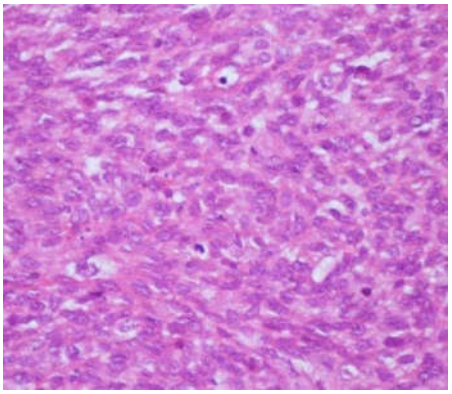

Figure 3: Mitotic Figures are present

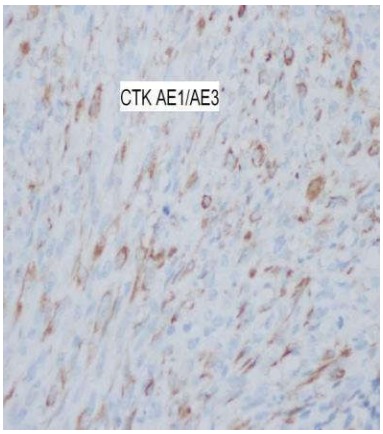

Figure 4: The tumor shows focal positive staining for cytokeratin in the cytoplasm of the tumor cells

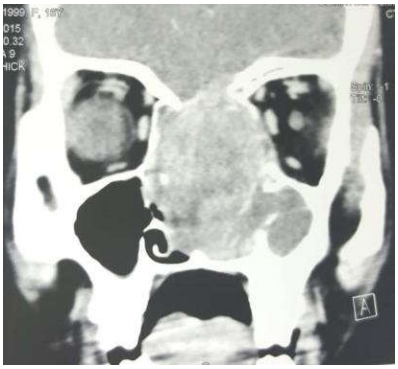

Figure 5 [Recurrence in the form of a left naso-ethmoidal tumor extended to the base of the skull, orbit, right nasal cavity and right ethmoidal sinus