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CLINICAL AND HISTOPATHOLOGICAL ASPECTS IN HEMANGIOPERICYTOMA

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Keywords: *type of soft-tissue tumour, malignant potential, hemangiopericytoma, pericytes, autosomal recessive transmission, age of 40, childhood, infiltrative and vegetative tumour formation, cavum*

Abstract: *Described as a type of soft-tissue tumour with malignant potential, hemangiopericytoma originates at the level of the pericytes, cells located in the walls of the capillaries. Although the etiology is not completely elucidated, it seems that the genetic factor plays an important role in the appearance of this type of tumour. Several cases with autosomal recessive transmission have been described in the literature. It is a formation that can occur at any age, but is more common after the age of 40. In fact, following multiple scientific studies, it has been concluded that the average age of affected people is somewhere around 45 years, while the survival rate at 10 years is 70%. Several cases have also been described in childhood (the percentage of occurrence at this age is between 5 and 10%). The sex incidence is equal. In what follows, I thought of presenting the case of a 90-year-old male person, diagnosed with an infiltrative and vegetative tumour formation located in the cavum, which turned out to be, after the anatomopathological examination, a hemangiopericytoma.*

INTRODUCTION

Hemangiopericytoma (1,2,3,4,5,6) is a vascular tumour (7) with malignant potential, fortunately, quite rare. The first case of hemangiopericytoma was described in the literature by Stout and Murray in 1942.(8) Hemangiopericytoma (9,10) develops from capillary vessels and is located more frequently on the skin of the head and extremities, in the subcutaneous cellular tissue but also in the skeletal muscles of the lower limbs. When located at the retroperitoneal level, this tumour may be accompanied by manifestations that include hydronephrosis, hydroureter, abdominal distension, or vomiting.(11) In a low percentage, this tumour formation can be highlighted at the synonasal level. It may be accompanied by nasopharyngeal bleeding or nasal obstruction. Although they are rare at this level, their prognosis is better because they tend to be less aggressive and do not metastasize. Macroscopically, it is presented as a unique formation, of variable dimensions, well (12) or very well delimited, surrounded by a well-vascularized pseudocapsule. Numerous vascular spaces are highlighted on the section surface, and the colour varies from gray to red-brown, with rapid development, which spreads to the brain and spine. A possible malignancy is determined by the presence of metastases or local recurrence. The treatment is exclusively surgical and consists in the total resection of the tumour formation. This type of treatment followed by radiation therapy (13) seems to be the most effective recovery method.

CASE REPORT

In this article, I set out to present the case of a 90-year-old person, without significant personal pathological history, who presents to the ENT service, complaining of respiratory difficulties. The patient is hospitalized for a progressive right nasal obstruction. Examination of the nasopharynx with palatal wave lift indicates the presence of a tumor formation with infiltrative character but also vegetative located in the cavum. The patient performs a CT examination of the nasopharynx which shows the presence of an isodense tissue formation with cervical muscles, which almost completely obstructs the lumen, with dimensions of 6/3 / 2.8 cm, intense and inhomogeneous iodophilic, the contrast uptake being better expressed in right half. The formation extends cranially in contact with the bilateral pharyngotmpanic tube, compresses the splenius capitis muscle, the lifting muscle of the palatine wave, the soft palate and also involves the posterior pole of the nasal cavities, being in contact with the middle nasal horns. Through the osteomeatalemeatal complexes it extends to the posterior ethmoidal cells, bilaterally.

This formation does not cause osteolysis of adjacent bone structures (including the nasal septum), but causes a slight hyperostosis of the right pterygoid plaque and sphenoid body. The presence of pathological latero-cervical lymphadenopathy is not observed.

The CT diagnosis formulated following the presence of these changes is: Nasopharyngeal tumour with loco-regional compressive effect. Subsequently, under local anesthesia, a biopsy of the tumour was performed, necessary to perform a histopathological examination. During the hospitalization, the patient underwent hemostatic, antibiotic and analgesic treatment, after which no acute complications appeared. It is discharged on request. About 3 months after discharge, the patient's evolution is good, even in the absence of surgery.

DISCUSSIONS

Macroscopically, 3 fragments were received with diameters between 2 and 4 mm, whitish-gray, elastic consistency, fully included. Microscopically, fragments of fibroconjunctival tissue (figure no. 1), partly lined with a respiratory epithelium, partially of a simple epithelium, could be

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highlighted that show a diffuse proliferation composed of relatively uniform cells in size (14), round-oval shape (15,16), with variable eosinophilic cytoplasm (17) (figure no. 2) and round nucleus located centrally, with poorly represented cell boundaries, with minimal atypia and rare mitosis.(18,19)

Figure no. 1. Optical microscopy image in hematoxylin-eosin staining, with 20x objective, what highlights the presence in a fibroconjunctival tissue (with red arrows) evenly of numerous cells of various shapes, with a disordered disposition (black arrows)

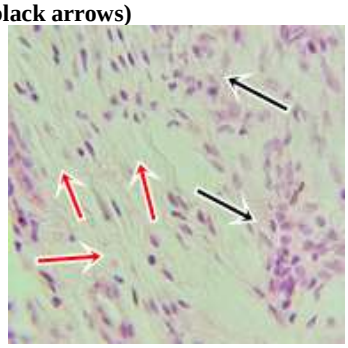


Figure no. 2. Optical microscopy image, in hematoxylin-eosin staining, with 10x objective, shows a proliferation of round cells arranged with approximately equal dimensions (this arrangement is observed throughout the microscopic field)

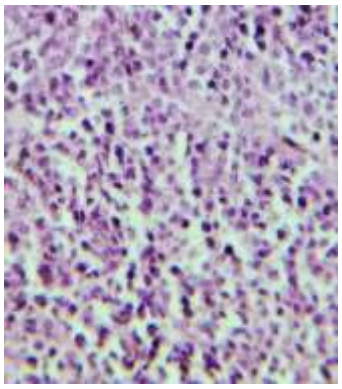
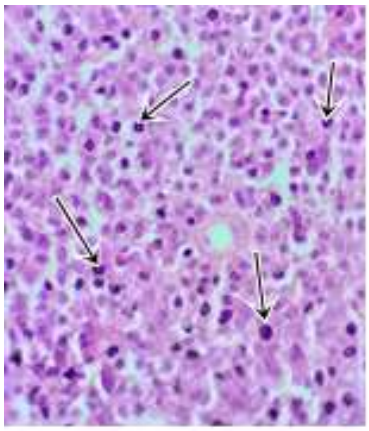


Figure no. 3. Optical microscopy image, hematoxylin-eosin staining, with 20x objective, shows a proliferation of round cells of approximately is equal size, with round nuclei (black arrows)



You can also see areas where fibroconjunctival tissue is richly represented, while cellularity is low (figure no. 4). The cells are located predominantly around the blood vessels with different caliber (figure no. 5).(20,21) Perivascular fibrosis phenomena can also be observed. Glandular elements are associated with various shapes and sizes, lined with a cubic epithelium, some with eosinophilic content, but also reduced fat component.

Figure no. 4. Optical microscopy image, in in hematoxylin-eosin staining, with 20x objective in which the presence of fibroconjunctival tissue (arrows) observed, abundant in quantity, but with reduced cellularity

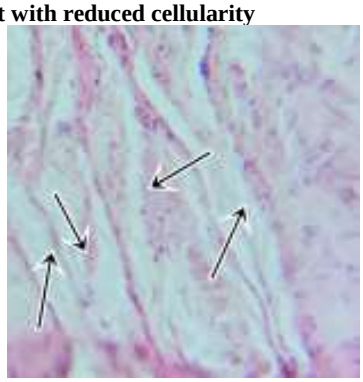
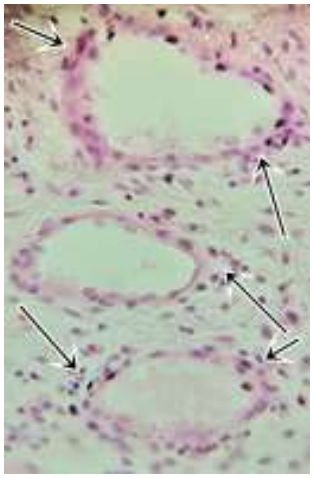


Figure no. 5. Optical microscopy image, in hematoxylin-eosin staining, with 20x objective, in which the arrangement of the cellularity around the blood vessels can be observed (arrows)



CONCLUSIONS

The ENT examination as well as especially the MDCT examination brought important information regarding the presence of symptoms (right nasal obstruction and epistaxis) and the location of the tumor formation. But the histopathological examination could lead us to a definite diagnosis, that of synonasal hemangiopericytoma. The anatomopathological examination was doubled by performing additional immunohistochemical tests (using certain antibodies) that revealed certain changes. These extremely valuable markers are used to differentiate a hemangiopericytoma and other types of malignant tumors or tumors with potential malignancy. In the present case, the following antibodies were analysed:

Vimentin - A nonspecific intracytoplasmic filamentary protein. Confirms the mesenchymal origin in certain tumours. It is well expressed in astrocytes and in developing neurons. It is positive in tumor proliferation.

Actin - Is a fibrillar protein. It is present either as a free monomer called G-actin (globular) or as part of a linear polymer microfilament called F-actin (filament), both of which are essential during cell division. It is positive in tumor proliferation.

Cromogranin - It is a secretory protein, negative in hemangiopericytoma. It is used to rule out the presence of neuroendocrine tumors (paraganglioma).

S100 - Negative in this case, is a useful marker in the case of tumors of the central nervous system (ependymomas, astrocytomas, oligodendrogliomas). Also, S-100 protein stains, with collagen IV, are particularly helpful for demonstrating Schwann cell differentiation in benign and malignant peripheral nerve sheath tumors.

Cytokeratin AE 1-3 - Negative in this situation, is a mixture of antibodies, used to highlight the presence of metastases. It is used to exclude the presence of epithelial tumors.

Cytokeratin MNF116 - Is a monoclonal antibody. It is used to rule out the presence of epithelial tumors. It is negative in this case.

Cytokeratin 5/6 - An important biomarker for different types of cancer (breast cancer and lung cancer). It is negative in this situation.

Cytokeratin 7 – Negative in this case, it is a low molecular weight cytokeratin. It is used to exclude the presence of a adenocarcinoma (lung, gastrointestinal).

Cytokeratin 20 - Similar to the previous one, it is a protein commonly found in various types of carcinomas (colon or urothelium adenocarcinomas). In hemangiopericytoma this antibody is negative.

CD3 - Is a protein complex and T cell co-receptor that is involved in activating both the cytotoxic T cell and T helper cells. It is negative in this case.

CD5 – Negative in this situation, it is a good immunohistochemical marker for T-cells, although not as sensitive as CD3.

CD10 - Common acute lymphoblastic leukemia antigen, it is negative in this case.

CD20 - Precious tumour marker in certain forms of lymphomas, negative in the case of hemangiopericytoma.

Bcl 2 – It is negative in this case.

HMB45 - Used in pathology as a marker for melanocytic tumors. Negative in this situation.

Ki67 - Is a nuclear protein associated with tumor cell proliferation and growth. Is an established prognostic indicator for the evaluation of biopsies from patients with cancer. It is an index of mitotic proliferation. In this case it is as a valuable prognostic biomarker for adult patients with hemangiopericytoma - 50%.

Due to the histopathological and immunohistochemical aspects listed above, we can make a diagnosis of synonasal hemangiopericytoma.

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