Case Report

A RARE CASE OF SCALP ANGIOSARCOMA IN A PATIENT WITH SUPERIORSAGITTAL SINUS MENINGIOMA

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ABSTRACT

Cutaneous angiosarcoma (CAS) is a rare but highly aggressive sarcoma of mesenchymal origin with a high mortality rate. The most affected sites of CAS are the scalp and facial skin. The 85-year-old patient, in follow-up for posterior parasagittal meningioma for several years, developed a single violaceous and ulcerated scalp lesion. The CT cerebral angio revealed, beyond the well-known meningioma, an area of bone rarefaction associated with ulcerated tissue infiltrating the skin and subcutaneous layers. After surgical biopsy, the histological examination documented a scalp angiosarcoma. The oncological treatment was not possible, given the age. The patient was referred to the plastic surgeon and underwent serial medication. The patient made a good functional recovery but died six months later due to a pulmonary embolism. This paper discusses the correlation between the scalp's angiosarcoma disease and the presence of the parasagittal meningioma.

KEYWORDS: *scalp angiosarcoma, sagittal sinus meningioma, head and neck tumor*

INTRODUCTION

Angiosarcomas of the head and neck represent about 15% of all head and neck sarcomas and 1% of all soft tissue sarcomas, most commonly arising on the scalp. Angiosarcomas originate from blood or lymphatic vessels and show a propensity for insidious local infiltration, for which they are considered aggressive tumors that recur both locally and distantly, with a risk of spreading through draining lymphatics. Given the rarity of this disease, there is limited evidence for its pathogenesis, primarily derived from case series. Angiosarcomas' aggressive behaviour causes estimated survival rates at 5 and 10 years are 34% and 14%, respectively.

There is considerable literature evidence supporting the hypothesis of the collateral veins pathway and vascular rearrangement forming in the presence of a superior sagittal sinus neoplasm. In this rare case, the authors suggest a cause/effect relationship between the superior sagittal sinus (SSS) meningioma with the superficial vessel remodeling and the scalp angiosarcoma.

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The rarity of this malignant tumor and its association with a neoplasm of the intracranial district make this case even more striking, as far as it has been reported. Furthermore, it may represent food for thought for studying angiosarcomas' biological and histological behaviour.

CASE PRESENTATION

An 85-year-old man was clinically and radiologically followed up since 2009 for a right-sided parietal para-sagittal extra-axial lesion, likely a meningioma (Fig. 1).

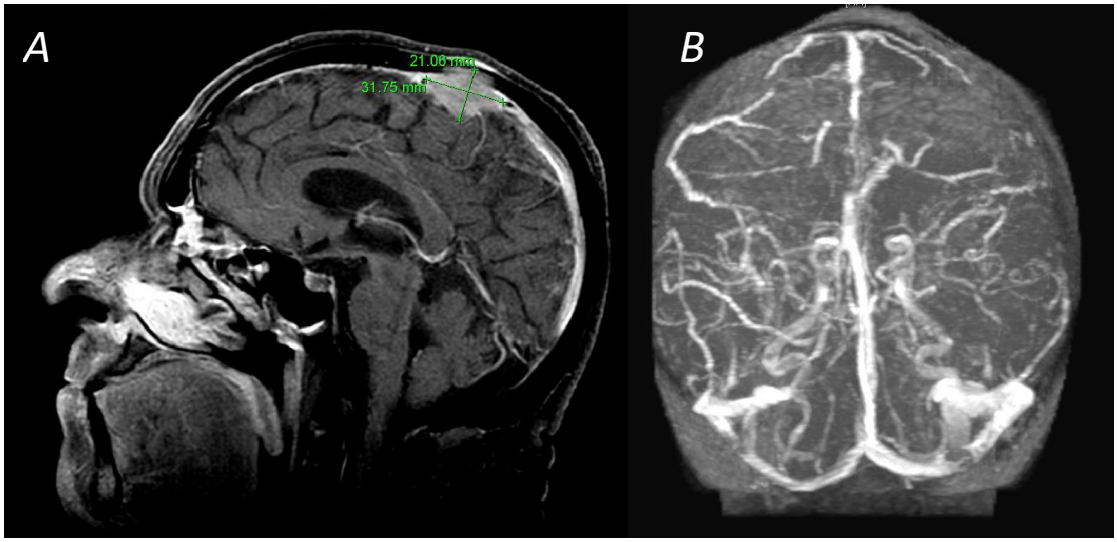

Fig. 1. *Enhanced MRI showing the superior sagittal sinus neoformation and angio-MRI highlighting the blood flow interruption in correspondence of the lesion.*

Because of the small dimensions and its slow growth over the years, a conservative conduct was adopted, even though a progressive and eventually complete sagittal sinus obliteration was observed after 3 years of diagnosis. Moreover, the neurological examination showed no disturbances. After an 11-year follow-up, cutaneous bleeding ulceration appeared on the left frontal area of the scalp (Fig. 2).

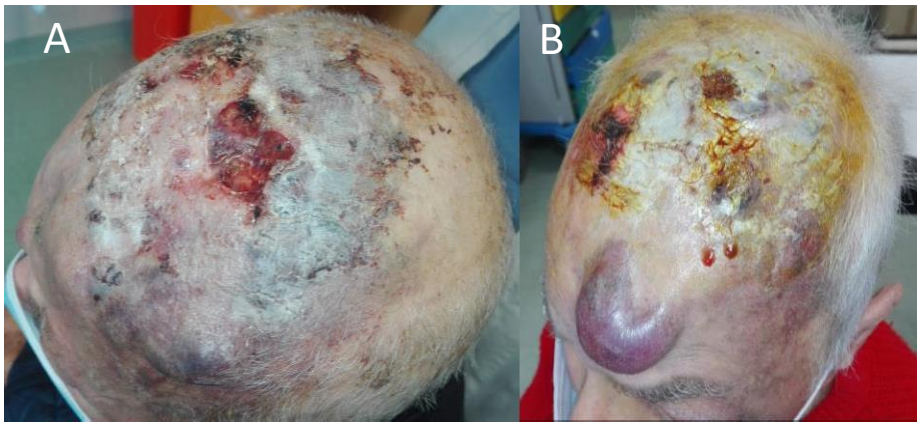

Fig. 2. *Ulcerated bleeding scalp lesion.*

Since he was under anticoagulant therapy for atrial fibrillation, anticoagulant therapy was suspended to gain better control of the bleeding and therapy with sodium heparin started. A new head CT scan with 1 mm 3D bone reconstructions revealed a considerable osteorarefaction area in correspondence with the known meningioma associated with a soft epicranial tissue infiltration (Fig. 3).

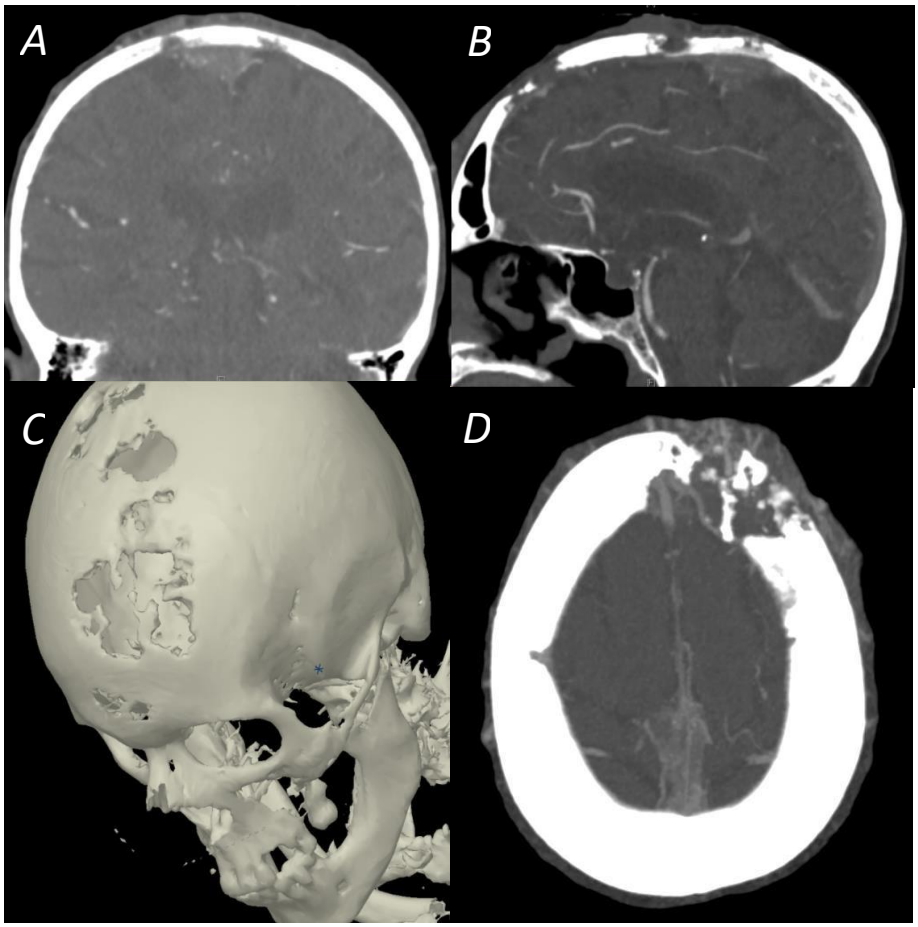

Fig. 3. *CT scan with bone windowing shows bony resorption areas.*

Considering the patient's global clinical conditions and comorbidities, no surgical indications were given, and the patient was referred to the plastic surgeon and underwent serial medications. A biopsy of the ulcerated tissue was performed, and the histological examination documented a scalp angiosarcoma. The tumor was composed of spindle cell proliferation with the formation of vascular solid areas, with positive immunohistochemical tests for CD34 and ERG negative for HHV8 (Fig 4).

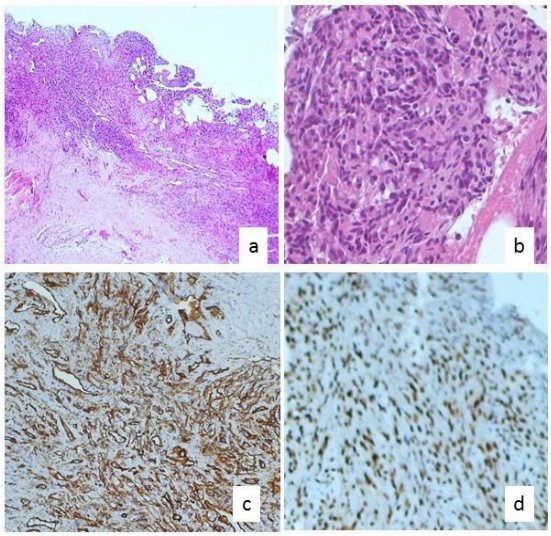

Fig. 4. *Tumor composed by spindle cell proliferation with formation vascular solid areas, with positive immunohistochemical tests for CD34 and ERG negative for HHV8*.

Radiotherapy was indicated to control the lesion size, but the poor patient's general conditions did not allow the treatment. The patient's overall survival was 6 months from the diagnosis, during which the patient experienced a good quality of life. In the last ten days, his clinical conditions suddenly became critical, causing a pulmonary embolism with rapid exits.

DISCUSSION

In literature, malignant tumors occurring on the scalp are reported as 1.4%-2% of all skin cancers, generally involving middle-aged and elderly individuals, and the differential diagnosis includes mainly basal cell and squamous cell carcinoma, malignant melanoma, and, most rarely, angiosarcoma.

Our study aims to find a correlation between the formation of the scalp collateral veins and their potential contribution to angiosarcoma's pathogenesis. When in the presence of a neoplasm infiltrating the superior sagittal sinus, a collateral venous circulation generates (to supply the cerebral territories otherwise lacking a venous drain and to bypass the obstruction site). The haemodynamic changes produce a global decrease of the oxygen partial pressure in the venous circulation to establish a hypoxemic micro-environment that may represent the starting point of a tumoral progression pathway. Maeda et al. reported that hypoxia enhanced angiosarcoma cells' proliferation, migration, and invasion ability (1). These findings suggested that the hypoxic tumor environment worsened the immune escape of angiosarcoma. Moreover, hypoxia increases angiopoietin-2 production, which results in abnormal angiogenesis in the presence of VEGF-A and vascular imbalance (2-4). Based on these experimental data using both clinical samples and cultured angiosarcoma cells, they enlightened the clinical importance of hypoxia in angiosarcoma.

Collateral scalp vein formation in the presence of a superior sagittal sinus neoplasm is a well-known phenomenon widely reported in the literature (5). Waga et al. (6) observed an abnormal filling of scalp veins in carotid angiograms in the following conditions: a) arteriovenous malformations (AVM) of the scalp; b) malignant tumors involving the scalp and skull; c) shunt blood into the dural venous sinus causing the pressure in the sinus to be increased (intracranial AVM and others); d) SSS occlusion with parasagittal meningiomas and venous sinus thrombosis (7, 8).

In scalp collateral veins, the pO_2 value is lower because of the venous engorgement produced by the small calibre vessels of the superficial scalp veins (temporal superficial vein, occipital vein, posterior auricular vein with final drain in the external jugular vein).

When the invasion leads to SSS stenosis or occlusion, venous collaterals are established via three known pathways: 1) collaterals connect the two ends of the occluded segment of the SSS; 2) collaterals bypass the occluded segment of the SSS but connect with a superficial venous system; 3) collateral bypass the occluded segment of the SSS but connect with a deep venous system.

Wollschaeger et al. (9) examined the rush of blood through dural infiltrating lesions (scalp basal cell carcinoma and para-falcine meningioma) with a selective external carotid angiography. They suggested that the relative stasis of the contrast material within the dural vessels could be explained by the growing resistance and encumbered redundance of capillaries within the lesion, whose peripheral vascular resistance resulted in decreased. Therefore, primarily dural lesion such a meningioma may determine prolongation and stasis of dural circulation.

Waga et al. reported 3 cases of parasagittal meningiomas occluding the SSS supplied by the scalpcollateral veins (6). The phlebogram of the internal carotid artery showed that veins placed anteriorly and posteriorly to the obstruction site drained mainly in a dilated vein in the posterior frontal region, descending in the pre-auricular region and finally in the retro-mandibular vein. The dilated scalp vein was a superficial temporal vein, receiving blood through emissary veins from the patent portion of the SSS. Even seen with the naked eye during surgery, they were markedly dilated and descended from the biparietal bony prominences down to the pre-auricular regions. The author describes an interesting case of a patient previously diagnosed of occipital region sarcoma, subsequently undergone surgery for a voluminous angioblastic meningioma of the occipital region infiltrating and occluding the SSS on its posterior third. The phlebograms showed several scalp veins connected with the SSS via emissary veins, all joined together and became a single vein, the superficial temporal vein, draining in the external jugular veins through the retro-mandibular vein.

In our case the scalp venous collateral circulation developed in presence of the SSS obstruction with relative blood flow stasis and hypoxia may have triggered the oncogenesis of the angiosarcoma. Our hypothesis should be investigated with an eventual confirmation by further biomolecular and oncological studies.

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