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Case Report

Sarcomatoid Carcinoma of the Orbit: A Rare Case Report

El Houssaine Boudra[1]*, Hajar Hanguir[1], Imane Gouzi[2], Ahmed Bennis[1], Nawal Hammas[2], Fouad Chraibi[1], Meriem Abdellaoui[1] and Idriss Benatiya Andaloussi[1]

[1]Department of Ophthalmology, University Hospital Hassan II, Sidi Mohamed Ben Abdellah University, Fez, Morocco

[2]Department of Biology, University Hospital Hassan II, Sidi Mohamed Ben Abdellah University, Fez, Morocco

***Corresponding Author:** El Houssaine Boudra, Department of Ophthalmology, University Hospital Hassan II, Sidi Mohamed Ben Abdellah University, Fez, Morocco.

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Abstract

Sarcomatoid carcinomas (SCs) are a variant of squamous cell carcinomas. They are extremely uncommon in the orbit. In the English literature, we reported Only 9 cases with orbital origin of this tumors. The prognosis of sarcomatoid carcinoma is extremely poor and the disease is usually clinically advanced at presentation. Currently, there is no consensus for the treatment of this variant malignant neoplasm. The treatment should be aggressive, and frequently yield poor results. We present a case of 42 years old male with orbital sarcomatoid carcinoma.

Keywords: Orbital; Sarcomatoid; Carcinoma; Case Report; Spindle Cell

Introduction

Sarcomatoid carcinoma (SC) is a rare variant of squamous cell carcinoma with sarcomatoid features. The localistion of this malignant neoplasm in diverse sites such as upper aerodigestive tract, genitourinary tract and uterus have been reported [1]. Orbital involvement is extremely rare, the clinical presentation is usually advanced and the treatment is generally aggressive.

Case History

A 42 years old male with Psychomotor delay since childhood. The symptoms go back for 4 months with the appearance of a rapidly progressive left orbital mass. On clinical examination of the left orbit had a exophytic orbital mass measured 7 cm of its large diameter from the base of the orbit invading all the eyeball and the eylids (Figure 1). The examination of the right eye as well as general examination does not find signs of remotely tumoral extending.

Figure 1: Clinical photograph showing tumor of the left orbit invading all the eyeball and the eyelids.

Orbital Computed tomography (CT) scan revealed a mass arising from Intra-orbital cavity with exophytic development outside the orbit involving the eyeball, optic nerve and ophthalmic artery as well as oculomotor muscles with the levator of the upper eyelid without visible bone lysis (Figure 2). The CT chest-abdomen-pelvis showed no secondary sites.

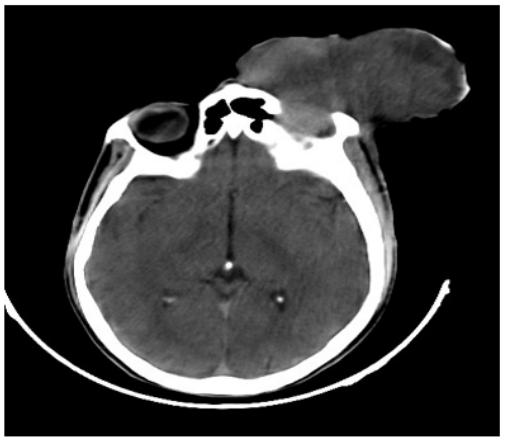

Figure 2: CT scan demonstrating mass arising from the left orbit with exophytic development outside the orbit involving the eyeball.

Biopsy of the mass showed a diffusely infiltrative neoplasm composed of cells rounded or spindled shape, with pleomorphic nuclei, strongly nucleolate in some area. The cytoplasm is abundant, well limited, sometimes clear and sometimes eosinophilic. Mitosis is frequent (Figure 3).

Figure 3: Photomicrograph showing undifferentiated malignant neoplasme proliferation composed of oval cells or polygonal, spindled shape in some area, with pleomorphic nuclei.

In immunohistochemical study The biomarkers Cytokeratin 7 (CK7), Cytokeratin 5/16(CK5/16) and vimentin reported positive. The tumor was negative for melanoma and Muscular markers. In the light of this results, a Sarcomatoid carcinoma was diagnosed.

The treatment suggested was orbital exenterating followed by the radiotherapy. After surgery the patient had decided not to proceed further treatment and was lost to follow-up.

Discussion

Sarcomatoid carcinoma is a type of malignant biphasic tumour with mixture of squamous cell and the sarcomatoid components. They are uncommon in the orbit. In 1899 Virchow reported the first case of carcinsarcoma with uterine location. The first case of orbital origin was reported in 1992. Using a PubMed search identified 9 cases of orbital carcinosarcoma [2-4].

Multiple hypotheses have suggested that Sarcomatoid carcinoma is derived from 2 distinct cell lineages [5]. More recent studies support a monoclonal epithelial cell origin with subsequent reprogramming of descendant cells, which results in the formation of mesenchymal cells [6]. In some previous series; radiation, trauma, tobacco use or alcohol consumption were reported as etiological factors [7].

The immunohistochemical study are needed for diagnosis of this variant carcinoma, and both epithelial and meschymal markers almost reported positive, most of these tumors stain with both epithelial and meschymal markers. The pancytokeratin is positive from 26% to 62% of cases [8,9]. Other common epithelial markers including EMA and p63 have been reported positive for some cases [9]. Vimentin; as mesenchymal-type markers, are positive in almost 100% of cases [8,9].

The treatment for sarcomatoid carcinoma is usually aggressive. The full resection with radiotherapy is mainstay of treatment and less often chemotherapy [3]. Owing to the rarity of the cases in the literature, there is no standard recommendations for the treatment of this tumor entity. Prakalapakorn., *et al*. [4] reported successful surgical resection in one patient with orbital carcinosarcoma with no re-currence reported 10 months after surgery. However, death occurred 13 months after surgical intervention in another patient. In one case reported by Ninan Mathew., *et al*. [3] the enucleation

was carried out with radiotherapy, The patient opted not to proceed with more aggressive surgical options or chemotherapy due to comorbid disease, general frailty, and risks of complications, a subsequent MRI showed stable disease.

The SC tend to pursue an aggressive course, characterized by recurrences and metastases after local treatment [5]. The prognosis is poorer with advanced patient age, and the occurrence of distant metastases [10].

Conclusion

The sarcomatoid carcinoma is a uncommon and aggressive tumor of the orbit, whose histogenesis is controversial, and it has, in most cases, a complex Histological characteristics. Although knowledge of the clinicoanatomical features of this tumor is necessary for appropriate management.

Statement of Ethics

All details of this case were reproduced after obtaining consent from the patient and the patient's next of kin. Any treatments undertaken by the patient were without deviation from accepted standards of care.

Disclosure Statement

The authors declare no actual or potential conflicts of interest.

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