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Cribriform adenoid cystic carcinoma (ACC) in the upper medial part of the left orbit presenting with exophthalmos – a case report

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ABSTRACT

Introduction: Orbital tumours in adults are the most common cause of unilateral, painless, non-inflammatory exophthalmos. Localization of the tumour in the oculomotor muscle or infiltration of the optic nerve causes more disturbing symptoms, such as the appearance of double vision – diplopia and/or decreased visual acuity.

The most common cause of orbital tumours in adults is cancer metastasis (more often occupying the left orbit). Tumours can also spread to the orbit from surrounding tissues such as the sinuses, the nasal part of the throat or from the lacrimal gland.

Adenocarcinoma of the cystic gland (ACC) is a rare, slow-growing malignant tumour arising from glandular tissue, usually appearing around the age of 50. ACC usually involves the salivary glands but may involve other structures such as the nasopharynx, maxillary alveolar process, orbit, nasal cavity or lacrimal gland.

Case report: A 53-year-old man presented to the hospital with visual disturbances and exophthalmos of the left eyeball. Magnetic resonance imaging showed the presence of a well-demarcated solid mass measuring 37 x 20 x 36 mm, located in the upper medial part of the left orbit, without features of diffusion restriction, with intense contrast enhancement. The lesion involved the intraosseous part of the left orbital roof with thinning of the cortical layer of the inner lamina of the frontal bone. The lesion protruded into the posterior situs on the left side. The tumour compressed the levator palpebrae superioris muscle and the superior rectus muscle and the distal part of the intraorbital optic nerve. Small chronic vascular foci were observed subcortically and in the periventricular white matter. There were also inflammatory changes of the cuneiform sinus and the mastoid process of the left temporal bone with the presence of fluid collections and a left deviated nasal septum.

Histopathological examination identified adenocarcinoma type cribriform. Access was obtained through a pteroidal-anterior craniotomy. The tumour was infiltrating and passing through the orbital roof. The tumour was dissected and removed from the septum, situs and cuneiform sinus. The postoperative course was uncomplicated, there was an improvement in visual acuity.

Discussion: The prognosis of patients with ACC is unfavourable (Bhayani et al. 2012; Jang et al. 2017; Ko et al. 2007; Xu et al. 2017). Metastases may occur even after many years and high mortality is usually due to intracranial dissemination or distant metastases. The difficulty in achieving complete surgical resection is due to the complex orbital anatomy, which leads to frequent local recurrence. The histopathological type of tumour has also been shown to have prognostic significance. In most cases, ACC does not occur as one pure histological type and classification is based on the dominant form. There are three histopathological types of ACC: cribriform (the most common, moderately differentiated), tubular (well-differentiated and with the best prognosis) and solid (poorly differentiated).

INTRODUCTION

Adenoid cystic carcinoma (ACC) belongs to the group of malignant neoplasms typically originating from the salivary glands (Ammad Ud Din and Shaikh, 2022). In total, it constitutes 10% of all salivary gland tumours (Dillon et al., 2016a). It is typically slow-growing compared to other neoplasms and tends to invade perivascularly as well as haematogenously disseminate to distant organs and is most commonly seen in older populations (Chummun et al., 2001). This kind of lesion constitutes only around 1% of orbital tumours, thus its occurrence in this location is particularly rare (Font, 1998). The onset of the illness is typically characterised by the indolent and relatively sluggish growth without any symptoms of the patient (Dillon et al., 2016b). However, the tumour has a tendency for local infiltration as well as perivascular proliferation thus the first sights of illness may be caused by the infiltration of nerves or other structures located around the lesion. (Khan et al., 2001). The consequences of oculomotor

involvement and infiltration of the optic nerve are more disturbing symptoms such as the appearance of double vision – diplopia and/or decreased visual acuity (Chen et al., 2017; Najem and Margolin, 2022). The spread of the tumour toward structures with reduced resistance within the orbit leads to compression of venous vessels and displacement of the eyeball toward the from the tumour to the space of least resistance, the eyelid crevice (Bradley, 2017). It is also associated with a high risk of both local recurrence and intracranial proliferation (Moskaluk, 2013). Moreover, the correlation between the histological structure of the tumour and the prognosis appears. Presently, assessing the probability of late recurrences is still difficult (Jaso and Malhotra, 2011). The most significant methods of primary treatment are surgical resection and radiotherapy, but the worth of adjuvant therapies remains as a contentious issue (Ishida et al., 2020a; Kokemueller et al. 2004). During the course of illness, patients in many cases develop local recurrence and metastases, specifically in the lungs, bones and liver (Papaspyrou et al., 2011). The cause of the development of this kind of neoplasm seems to be impossible to indicate (Coca-Pelaz et al., 2015). Orbital tumours, despite the fact that they are a very rare condition and not a significantly epidemiological problem, are nevertheless the most common cause of unilateral, painless, non-inflammatory exophthalmos of the eyeball in adults. In the early stages of the disease, lesions that are mild and slow-growing such can be very subtle and often underestimated (Ishida et al., 2020b). Limited data are available on predisposing risk factors and the management of patients with advanced disease due to its rarity (Ouyang et al., 2017; Singaraju et al., 2022). Pathophysiology of ACC is a scarcely investigated area due to the rarity of the condition (Chae et al., 2015).

The most common cause of orbital tumours among adults is metastatic cancerous tumours. Primary tumours can occupy the breast (most commonly), lungs, urinary and genital tract (especially the prostate gland) (Lukšić et al., 2016). Moreover, tumours spreading to the orbit from the tissues surrounding, such as the sinuses of the nose (most often), the cranial cavity, and the eyelids. Other tumours of the orbit originate from vascular tissue, lymphoid tissue, nerve tissue and the lacrimal gland (Xiao et al., 2019). Stasis with swelling and vasodilation of the conjunctiva are also observed. Orbital tumours rarely provide the pain, except in the case of lesions that rapidly increase that may be the cause of the late diagnosis (Składzień J., 2000).

CASE REPORT

29th of January in 2018 year, 53-years old patient reported to the Department of the Ophthalmology in University Clinical Hospital in Zielona Góra because of the isolated exophthalmos of the left eyeball and visual acuity impairment. The patient reports that the complaints gradually increased over time until the patient's functioning became compromised. In the anamnesis the Diabetes Melitus and infected tooth decay. In addition, the patient reports severe pain in his lower extremities. He has recently been treated with metformin, methylprednisolone and amoxicillin. Due to the unilateral character of the symptoms and an absence of underlying disease the Magnetic Resonance Imaging has been ordered. The examination performed on 5th of January revealed the presence of a well-demarcated solid mass measuring 37 x 20 x 36 mm, located in the upper medial part of the left orbit, without features of diffusion restriction, with intense contrast enhancement. The lesion involves the medial portion of the ceiling of the left orbit with thinning of the cortical layer of the inner lamina of the frontal bone. The mass protrudes into the posterior situs on the left side. The tumour compresses the upper eyelid levator muscle, as well as the superior rectus muscle and the distal part of the intraorbital optic nerve. Small chronic vascular foci subcortical and in the periventricular white matter. Inflammatory lesions of the cuneiform sinus and the mastoid process of the mastoid process of the left temporal bone with the presence of fluid collections. Left convex curvature of the nasal septum. Trace of fluid in the alveolar lobe of the left maxillary sinus. A biopsy of the lesion was recommended, and histopathological examination of the retrieved material allowed a diagnosis of adenocarcinoma of the lacrimal gland type. The results were suspicious for adenocarcinoma of the lacrimal gland or from the mucosa of the paranasal sinuses; however, metastasis could not be entirely ruled out. Infiltration of nearby blood and lymphatic vessels was notable. The cribriform variant is the most common and consists of lobules with circular pools of mucin.

Staining for mucin (+/-) positive in the cytoplasm of some tumour cells. The final diagnosis was the adenocarcinoma, sieve type. It was decided to implement surgical treatment, a complete resection of the lesion along with a margin of healthy tissue. Operative access to the tumour was difficult, as the tumour infiltrated and passed through the roof of the orbit. Surgical treatment included a pteroidal craniotomy. At first, the tumour was separated from the septum, situs and afterwards the parts of the lesion were removed from the wedge sinus. The postoperative course was uncomplicated, moreover after the procedure the complaints of the patient disappeared and visual acuity was completely restored.

DISCUSSION

The role of the surgical treatment and postoperative radiotherapy seems to be crucial both in the process of restoring the patient's function and preventing or delaying tumour recurrence. However, strict follow-up after surgery should be implemented to assess for both local recurrence and radiation related complications (Esmaeli et al. 2006). In this case, due to the location of the lesion and the infiltration of the roof of the orbit, the resection was particularly difficult. Adenoma cystic carcinoma as the primary orbital tumours in the absence of lacrimal gland involvement is particularly rare. The absence of neoplastic cells in the lacrimal gland does not rule out the diagnosis of ACC in the assessment of an orbital tumour (Cantù, 2021). Given the aggressive nature of this neoplasm with a propensity for recurrence, intracranial dissemination and late distant metastasis, the diagnosis of adenoma cystic carcinoma should be considered in every case of the occurrence of the mass in the orbit (Venkitaraman et al. 2008).

CONCLUSION

The patient underwent the operation very well and the result was satisfactory. Bodily functions were restored and he regained his visual acuity. The above data seem to speak in favour of a timely surgery. Which can be, as shown by the case described by us, even at a very advanced stage. It should be remembered that tumours in this location are most often metastases and should be excluded first. Whenever possible, it is important to take surgical action which, together with rapid rehabilitation, allows the patient to regain his pre-disease condition and function. The role of surgical treatment and post-op radio-therapy appears to be crucial both in restoring the patient's function and in preventing or delaying tumour recurrence. However, close follow-up after surgery should be implemented to assess both local recurrence and radiation complications Despite the difficulty of the tumour location in the present case – infiltration of the orbital roof – the patient was successfully restored. This is an encouraging sign for neurosurgery in similar cases. However, it should be borne in mind that decisions should always be made on an individual basis with regard to the patient's personal predisposition. However, as suggested by (Venkitaraman et al., 2008) given the aggressive nature of ACC with its propensity for recurrence, intracranial spread and late distant metastasis, the diagnosis of adenoma cystic carcinoma should be considered in every case of an orbital mass and treated surgically (Ramakrishna et al., 2016; Vidović Juras et al., 2019).

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