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Niezróżnicowany rak zatokowo-nosowy – rzadka przyczyna wytrzeszczu oczu u starszych osób

Sinonasal undifferentiated carcinoma – a rare cause of proptosis in elderly

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Streszczenie

Niezróżnicowany rak zatokowo-nosowy to rzadki i niezwykle agresywny nowotwór przewodu zatokowo-nosowego. Charakteryzuje się szybkim i destrukcyjnym wzrostem. W niniejszej pracy przedstawiono przypadek 71-letniego mężczyzny pochodzenia malajskiego z utrzymującym się od miesiąca, postępującym wytrzeszczem lewego oka, któremu towarzyszyły lewostronne ograniczenie widzenia, okresowe podwójne widzenie oraz brak powonienia. Ostatecznie u pacjenta rozpoznano niezróżnicowanego raka zatokowo-nosowego. Po serii zabiegów radioterapii uzyskano poprawę stanu zdrowia chorego. Niniejszy opis przypadku wskazuje na konieczność uwzględnienia diagnozy niezróżnicowanego raka zatokowo-nosowego u osoby w podeszłym wieku, u której występują wytrzeszcz oczu oraz objawy nosowe.

Słowa kluczowe: wytrzeszcz oczu, niezróżnicowany rak zatokowo-nosowy

Abstract

Sinonasal undifferentiated carcinoma is a rare and extremely aggressive sinonasal tract malignancy. It is a rapidly enlarging tumour and a highly destructive lesion. We hereby report a 71-year-old Malay male with progressive left proptosis for one month, associated with reduced vision in the left eye, intermittent diplopia and anosmia. He was later diagnosed as having sinonasal undifferentiated carcinoma. The patient subsequently underwent a series of radiotherapy and showed improvement following treatment. This case was reported to highlight the importance identifying this entity and emphasise that sinonasal undifferentiated carcinoma should be considered in an elderly patient with proptosis associated with nasal symptoms.

Keywords: proptosis, sinonasal undifferentiated carcinoma

INTRODUCTION

Proptosis is described as an abnormal protrusion of the eyeball, reflecting an increase in orbital volume[1]. The causes of unilateral proptosis include trauma, endocrine ophthalmopathy, inflammatory disease like orbital cellulitis, tumour, including orbital haemangioma, malignant lymphomas and metastatic malignant tumours[1,2]. Sinonasal undifferentiated carcinoma (SNUC) is a rare and extremely aggressive malignancy. It is a rapidly enlarging tumour and a highly destructive lesion. SNUC often presents with nasal obstruction, nasal discharge, epistaxis, facial pain, proptosis, diplopia and diminution of vision[3].

CASE REPORT

A 71-year-old Malay male with underlying bronchial asthma presented with progressive left proptosis for one month. It was associated with reduced vision in the left eye, intermittent diplopia and anosmia. Ophthalmic examination showed visual acuity of 6/18 in the left eye and 6/9 in the right eye. Left eye examination showed axial proptosis associated with swelling of the left nasal bridge. There was restriction of ocular movement in all gazes as well. The anterior and posterior segment of the left eye was unremarkable with normal intraocular pressure. The right eye examination was normal. Other cranial nerve function was intact.

Nasoendoscopy assessment by an otorhinolaryngology team showed a mass filling the nasal cavity that pushed the septum to the right side. Biopsy of the mass was taken during nasoendoscopy procedure. Computed tomography (CT) showed a heterogeneously enhancing mass in the nasal cavity eroding the adjacent bone and extending into the adjacent maxillary, ethmoidal and frontal sinuses. It also extend into the bilateral extraconal orbital cavity, more on the left and caused proptosis of the left eye globe (Fig. 1). Magnetic resonance imaging (MRI) showed that this sinonasal mass, which measured 6.3 × 7 × 6.5 cm, extended into the anterior cranial fossa superoanteriorly, causing mass effect and oedema of the overlying frontal lobe. There were also involvement of the left medial rectus and inferior oblique extraocular muscle. The rest of the orbital content and the brain were preserved. There was also extension into the cavernous sinus.

Histopathological examination revealed features of undifferentiated carcinoma. Immunohistochemistry showed intracytoplasmic keratinisation in some cells. The tumour cells are strongly positive for cytokeratin 7 (CK 7), pan-cytokeratin (AE1/AE3) and tumour marker P16, and 50% of cells were positive for tumour marker P63. Thus, the final diagnosis of SNUC was made. The patient was scheduled for chemoradiotherapy. However, he refused chemotherapy and opted for radiotherapy only. He underwent 35 cycles of radiotherapy (dose 70 Gy). Repeated CT scan two months after completion of radiotherapy showed reduction of mass

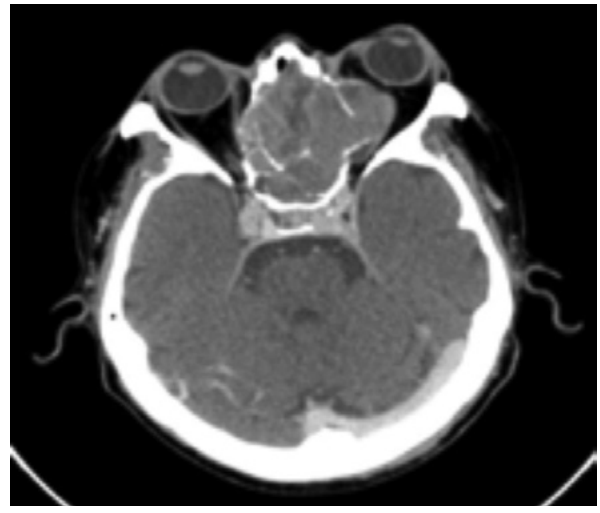

Fig. 1. A CT scan showed a mass in the nasal cavity, extending to the left orbital cavity and causing marked proptosis of the left eye

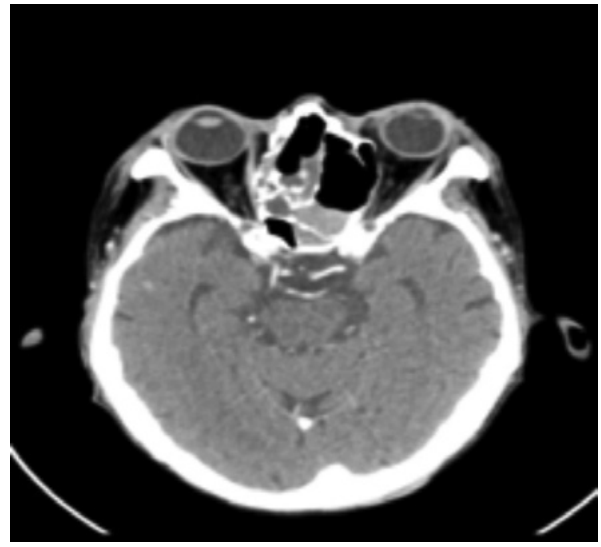

Fig. 2. A CT scan post radiotherapy shows reduction of mass within the nasal cavity. Proptosis is reduced

size with resolved previous left eye proptosis and frontal white matter oedema (Fig. 2). During his recent follow-up, which was seven months after the presentation, the proptosis had resolved; however, there was still diminution of visual acuity and extraocular muscle restriction over the left eye. He is currently on 6-monthly follow-up.

DISCUSSION

Nasal cavity and paranasal sinuses malignancies account for <3% of head and neck malignancies. SNUC represents 0.2–0.8% of all malignancies[3]. Frierson et al. (1986) described it as aggressive neoplasm that was clinico-pathologically distinct from other poorly differentiated malignancies of the nasal cavity and sinuses[acc. to 4]. SNUC is predominantly more common in man than women (3:1 ratio), with the

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commonest age group in the fifth decade of life[5,6]. The aetiology of SNUC is still unclear. Risk factors such as smoking, history of radiation therapy and occupational exposure to coal, nickel and chrome have been postulated. However, unlike nasopharyngeal carcinoma, SNUC showed no association with Epstein–Barr infection[7].

Patients commonly presented with nasal obstruction and discharge, epistaxis, proptosis, diplopia and reduced vision. However, there was reported a case of SNUC presenting as metastatic spinal cord compression and frontal-ethmoid sinus swelling[8,9]. SNUC is a rapidly growing malignancy, involving multiple paranasal sinuses, commonly anterior nasal cavity and ethmoids[5]. It causes marked destruction of the sinus wall that leads to disease extension into the orbital cavity, intracranial cavity and nasopharynx[5]. Since early symptoms of SNUC are similar to those found in patients with benign sinus disease, it causes delay in seeking treatment. Therefore, SNUC patients usually present at advance stage at the time of diagnosis.

CT and MRI of SNUC show expansile, heterogeneous mass lesion with significant bone destruction and invasion of adjacent structures, including the anterior cranial fossa, adjacent paranasal sinuses and the orbit[10]. The involvement of the sinonasal tract structure is more anterior, including ethmoids as compared to nasopharyngeal carcinoma (NPC)[3,5]. Histologically, SNUC originates from Schneiderian epithelium which line the sinonasal tract. Immunohistochemical analysis is extremely helpful to differentiate SNUC from other undifferentiated malignancies, such as olfactory neuroblastoma and undifferentiated NPC. SNUC expresses simple epithelial type cytokeratine (CK 7, CK 8, CK 17) and epithelial membrane antigen (EMA) with variable reactivity towards neurospecific enolase (NSE), tumour marker p53, chromogranin and synaptophysin[4,11]. Most of SNUC malignant cells show no immunoreactivity towards S-100 protein and all are negative for vimentin[5].

Treatment generally involves surgical resection of the tumour. Craniofacial resection with maxillectomy, orbital exenteration and craniotomy has become the most common surgical approach[3]. Craniofacial resection with tumour-free margin and orbital exenteration in orbital involvement are positively related to tumour-free survival status[12]. However, patients with SNUC have a high rate of both local-regional recurrence and distant metastases. Furthermore, the complex anatomical structure of the head and neck make the complete removal of the tumour with wide margin not always possible.

Aggressive multimodality therapy including surgical resection and chemoradiation therapy became the current option of treatment[11]. Chemotherapeutic agents such as cisplatin, carboplatin, etoposide, doxorubicin, cyclophosphamide, vincristine, mitomycin-c and 5-fluorouracil had been used to treat SNUC[12]. Radiotherapy plays an important part of a comprehensive treatment as it can reduce local recurrences. It also improves the quality of life in patients with debilitating bony metastases by providing pain relief, preserving organ function, skeletal integrity and for palliative therapy[3]. Despite multimodality treatment of SNUC, the prognosis remains poor. Overall 5-year survival for SNUC is estimated at 22%[13].

CONCLUSIONS

SNUC should be considered in a patient with proptosis associated with constituent nasal symptoms. This may prompt initiation of diagnostic and therapeutic approach. It is important to identify this entity, especially in the early stage as an early intervention may result in higher possibility of survival and good outcomes.

Conflict of interest

The authors do not report any financial or personal connections with other persons or organizations, which might negatively affect the content of this publication and/or claim authorship rights to this publication.

Piśmiennictwo

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