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Atypical invasive secondary meningioma with extensive intracranial invasion: A case report

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ABSTRACT

Invasive and metastatic version of meningioma as an atypical type of the tumor has been rarely described in the literatures. Recurrent secondary meningioma invaded to pre-orbital soft and hard tissues can be found in extremely rare cases. We described a case of atypical invasive meningioma invaded to deep orbital soft tissue due to recurrence of the primary tumor previously treated by surgical removing and chemoradiotherapy. In this case, invasion to orbit, paranasal sinuses and masticator, infratemporal and buccal spaces leading to destruction of adjacent bony structures and mass engulfing the right globe resulting in considerable proptosis was the prominent feature.

Keywords: meningioma, invasive, secondary

INTRODUCTION

Meningioma is a partially common brain tumor with a prominence among women frequently middle or later adult life and consisting about 25 to 30 percent of all primary intracranial neoplasms [1]. This tumor commonly arises in proximity to meninges, however in rare conditions, its invasion to adjacent tissues and organ such as has been also reported different meningeal layers, brain vascular bed, and even skull has been also reported [2]. Although such invasive behavior may be remained asymptomatic, over time, the possibility of serious damage to organs and even a threat to the patient's life can be expected [3]. Moreover, the risk of tumor recurrence may be raised due to invasive growth of the tumor. Herein, we describe a case of atypical invasive meningioma invaded to deep orbital soft tissue due to recurrence of the primary tumor.

CASE PRESENTATION

The patient was 56-year-old women with the known case of meningioma that scheduled for chemotherapy and radiotherapy about 2 years ago leading clinical improvement, however she referred again with the complaints of headache and severe exophthalmia. In medical history, the patient was hypertensive with the controlled status at the time of referring (systolic/diastolic blood pressures of 140/90 mmHg) without other comorbidities. She was scheduled for CT scanning that revealed a huge, extra-axial, avidly enhancing mass in anterior and middle cranial fossa on the right side with invasion to orbit, paranasal sinuses and masticator, infratemporal and buccal spaces leading to destruction of adjacent bony structures. The mass engulfed the right globe resulting in considerable proptosis. The mass demonstrated restricted-diffusion. Cavernous and supraclinoid segments of right internal carotid artery were encased by the mass (Figure 1). The patient was candidate for craniotomy. In macroscopical observation, it was revealed a white-gray bloody mass sized 7.5 ×6.0 × 4.0 cm^3 adjacent to optic nerve with involving orbital soft tissue. The tumor was closed to eye globe without involvement of intraocular component, but extended to deep orbital tissue including muscle bundles. A sample of the mass was sent to laboratory for pathological assessment. In this regard, fragments of hypercellular neoplastic tissue were found with eosinophilic cytoplasm, ve-

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sicular atypical nuclei and occasional macro-nuclear nucleoli with mitotic figures about 4 to 5 numbers in 10 HPF but without any evidence of necrosis indicating atypical meningioma, the result of the recurrence of primary malignant tumor (WHO grade of II) (Figure 2). In immunohistochemical assessment, the tumor sample was positive for P53 (in 20% of tumor cells), Epithelial membrane antigen (EMA), and Ki67 (about 30 to 40% of tumor cells) (Figure 3).

DISCUSSION

Although meningioma is the most common extra-axial intracranial neoplasms, but the malignant mass with invasion and metastatic potential (higher WHO grades) are less common [4]. Due to the compressive effects of the mass and the possibility of invasion of the surrounding hard and soft tissues, the clinical manifestations of this invasion will be visible in the final stages [5]. Any suspicion to the presence of invasive meningioma should lead to imaging assessment. In brain CT scanning, an intra-axial or extra-axial hyperdense vascular mass occasionally with bone destruction and peritumoral edema can be revealed [6]. In post-contrast imaging, involving the skull and scalp may be appeared. The final assessment by angiography indicates intense vascular stain; however, for definitive diagnosis of tumor, histological assessment is essential. In pathological evaluation, 4 to 19 mitotic figures in 10 HPF can be the prominent figure with increased cellularity, tumor cells with high nuclear to cytoplasmic ratio, large nucleoli, loss of lobular architecture, or even geographic areas of necrosis [7,8]. In differential diagnosis, typical meningioma, dural metastasis, lymphoma, or Ewing sarcoma should be considered [8]. As elective treatment approach, gross total or subtotal resection followed by adjunctive radiation therapy should be considered [9].

As compared to typical meningioma, atypical meningioma is rare with poorer prognosis and high relapse rate higher than 25%. The 5-year survival rate in those suffering atypical meningioma has been estimated to be less than 90% with a 5-year recurrence rate of higher than 50%. In our case, a recurrent atypical meningioma was described with high extension to adjacent orbital tissues leading final diagnosis of invasive meningioma, grade II according to the WHO classification. Various cases

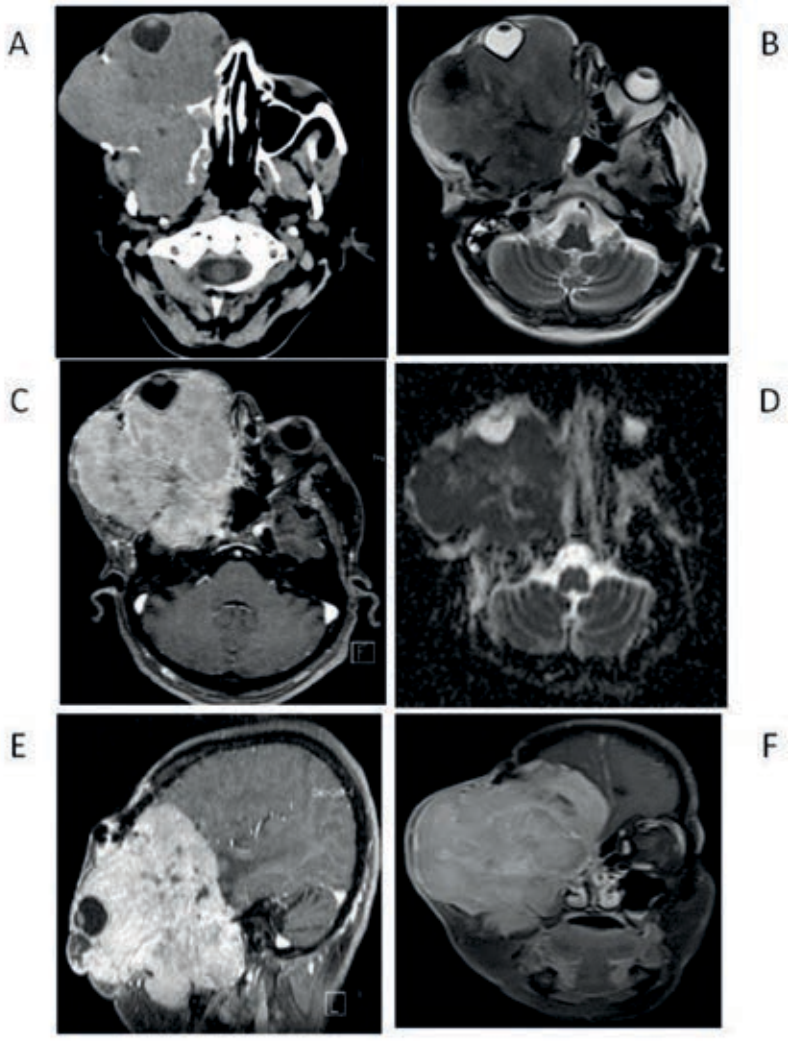

FIGURE 1. a) Axial non-contrast CT scan b) axial T2-weighted image c) axial T1-weighted image with gadolinium d) axial apparent diffusion coefficient (ADC) image e, f) sagittal and coronal T1-weighted images with gadolinium. There is a huge, extra-axial, avidly enhancing mass in anterior and middle cranial fossae on the right side with invasion to orbit, paranasal sinuses and masticator, infratemporal and buccal spaces and is leading to destruction of adjacent bony structures. The mass engulfs the right globe and is resulting in considerable proptosis. The mass demonstrates restricted-diffusion (d). Cavernous and supraclinoid segments of right internal carotid artery are encased by the mass

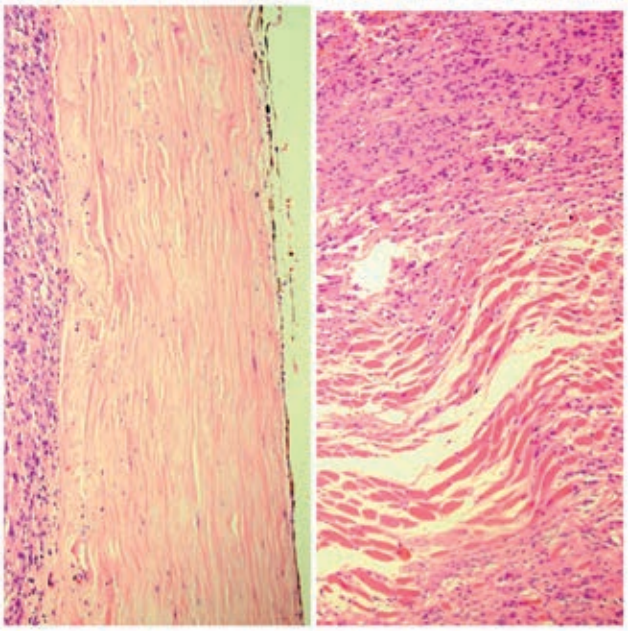

FIGURE 2. Histologic examination showed invasive atypical meningioma invades to optic nerve (left) and muscle bundles (right), H&E x 200

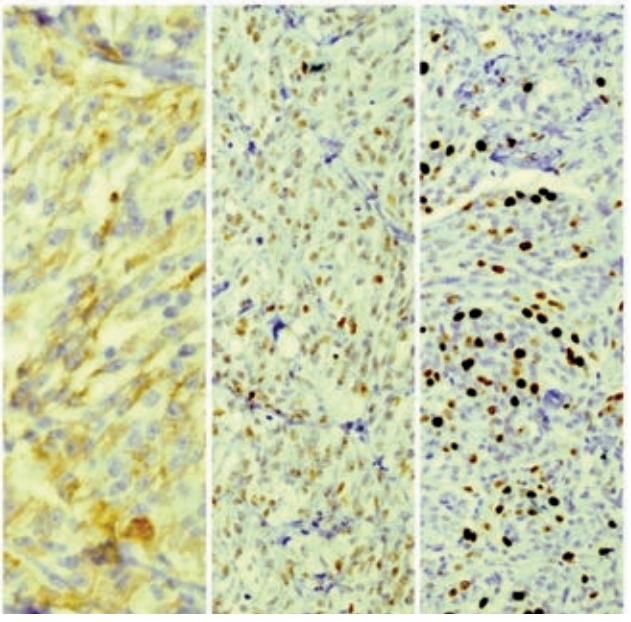

FIGURE 3. Immunohistochemical staining showed Positive epithelial membrane antigen (left), P53 staining (middle) and Ki-67 staining about 30% (right)

have been presented in the literature with the different invasive behaviors of such tumor. As described by Mrinal Matish et al. [10], an invasive meningioma with cystic intracranial neoplasm was described invaded to the left temporal fossa extending into the middle and external ear. In another case presented by Leison Maharjan [11], extracranial atypical meningioma of sinonasal tract was presented. Also, a case of primary atypical meningioma of the sinonasal Tract was described by Fechtner et al [12]. Ayşe Dolar Bilge et al. [13] also described their case of an atypical optic nerve sheath meningioma in a child that tumor size and visual acuity remained stable in the six months after treatment. What sets our case apart from the others described is that the case has been a recurrence of a tumor caused by a primary meningioma tumor with extensive invasion of the tissues around the eye. What is important in such cases is to diagnose the history of the primary tumor, make an accurate histopathological assessment, accurately track how it spreads or affects nearby tissues, and quickly determine a treatment approach to prevent adverse events due to tumor progression and tissue destruction.

CONCLUSION

Meningioma is common brain tumor with benign behavior and may be remained asymptomatic but seldom invades surrounding structures. Also, the risk of tumor recurrence may be raised due to invasive growth of the tumor. So, clinicians should be considered tumor's potential and quickly treat to prevent morbidity due to tumor extension.

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