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A CASE OF MULTIPLE INTRACRANIAL MENINGIOMAS

Medicine

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ABSTRACT

Meningiomas are the most common primary brain tumors in adults. Multiple meningiomas (MMs) or meningiomatosis are defined by the presence of at least two lesions that appear simultaneously or not, at different intracranial locations, without the association of neurofibromatosis. They present 1–9 % of meningiomas with a female predominance. The occurrence of multiple meningiomas is not clear. There are two main hypotheses for their development, one that supports the independent evolution of these tumors and the other, completely opposite, that suggests the propagation of tumor cells of a unique clone transformation, through cerebrospinal fluid. We present a case of a 60-year-old male patient who presented to the emergency department with weakness of right upper and lower limb. The patient was diagnosed with multiple meningiomas based on the magnetic resonance imaging of the brain. Multiple meningiomas is extremely rare in males.

KEYWORDS

Multiple Meningiomas, Vasogenic Edema, Hemiparesis.

BACKGROUND

Meningiomas are benign tumors arising from arachnoidal cap cells of the leptomeninges. The incidence of meningioma is 5.5 per 100000 people per year and accounts for nearly 13-26% of all the primary intracranial neoplasms in adults (1,2). Cushing and Eisenhardt (3) in 1938 were the first to coin the term meningioma. They defined multiple meningiomas as "at least two spatially separated meningiomas in a patient without signs of neurofibromatosis" (3).

Meningiomas are extra-parenchymal, and benign tumors of central nervous system that usually present with symptoms resulting from mass effect. Therefore, clinical presentation usually depends on the location of the tumor producing neurological signs and symptoms, epileptic fits or symptoms of raised intracranial pressure. Many are asymptomatic. However, they may contribute to significant surrounding edema, probably associated with their cellular morphology and capability of releasing vasoactive substances. Interestingly, the severity of edema is not related to the size of the tumor and may cause life-threatening increase in intracranial pressure (4,5).

Radiologic imaging is the main method for diagnosis and evaluation of meningioma. Computed tomography (CT) and magnetic resonance imaging (MRI) are preferred modalities. Proton magnetic resonance spectroscopy is a noninvasive technique that enables further assists and improvements in diagnosis of brain lesions, including meningiomas (6-8). Surgical resection is the standard treatment for meningiomas and endovascular embolization is usually performed before surgery to reduce tumor vascularity. Radiation therapy and hormonal therapy are indicated in cases of inoperability (9).

CASE REPORT

A 60-year-old male patient was brought to the emergency department with a chief complaint of weakness of right upper and lower limb for the past day. There was no history of headache, seizures, vomiting, fever and trauma. The patient was conscious and coherent. Pulse rate was 92 beats per minute, temperature was 98.4-degree Fahrenheit, respiratory rate was 15 breaths per minute and the blood pressure was 160/90 mm of mercury. On examination, the patient was aphasic and had a power of 3/5 in right upper and lower limb. The power was 5/5 in left upper and lower limb. There was no evidence of neurocutaneous markers. The emergency CT scan revealed an irregular area of hypodensity suggesting vasogenic edema in the left frontal area (Fig. 1) and multiple small areas of hyperdensity in the high parietal and occipital regions (Fig. 2).

FIGURE 1

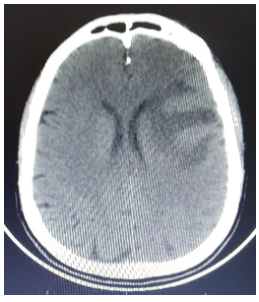
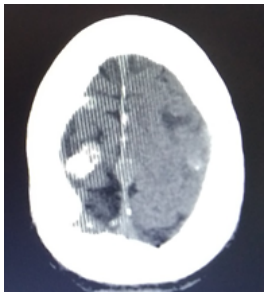


FIGURE 2



Fundoscopy revealed papilledema. We started a short course of steroids and intravenous 20% mannitol for reducing intracranial pressure (ICP). Meanwhile we obtained a contrast enhanced magnetic resonance image (CE MRI) of brain for further evaluation of underlying pathology. CE MRI of brain revealed the presence of 4.2 x 3 cm well defined dural based extra axial mass lesion with peri lesional edema in the left frontal convexity, which showed subtle diffusion restriction. Minimal effacement of adjacent lateral ventricle was noted with minimal midline shift towards right by 2mm. The lesion showed intense homogeneous enhancement on intravenous contrast. Magnetic resonance spectroscopy (MRS) of the lesion showed reduced N-acetyl-aspartate (NAA) levels and choline peak. There were similar lesions in the right frontal convexity, parietal convexity, anterior inter hemisphere fissure and left occipital convexity. These findings were suggestive of multiple meningiomas.

The patient improved symptomatically over the next three days. The patient left against medical advice while neurosurgery consultation was being made.

DISCUSSION

Meningiomas are tumors arising from arachnoidal cells, granulations, stroma of the perivascular spaces, and in the choroid plexus, corresponding to nearly 20% of all intracranial tumors (10). The term multiple intracranial meningiomas is used only when two or more meningiomas occur either simultaneously or sequentially in different locations (11). Multiple meningiomas constitute 1-2% of meningioma cases. With the advent of MRI, the incidence of multiple meningiomas is reported to be even higher because MRI detects even small tumors which are located in posterior fossa, skull base, and higher vertex area. (12)

Kyoi et al. (13) encountered two patients with multiple meningiomas at their clinic. Locatelli et al. (14) reported 10 cases of multiple meningiomas in a series of 227 intracranial meningiomas from 1977 - 1984. In this particular series, all the patients were female.

Domenicucci et al. (15) reported 14 cases of multiple intracranial meningiomas representing 1% of all meningiomas operated on at their hospital over a period of 35 years. Gelabert-Gonzalez et al. (16) reported 13 cases of multiple intracranial meningiomas, consecutively operated on at their hospital between 1983 and 2003. All those patients were studied with CT and a few with MRI, and all those patients showed no features of von Recklinghausen disease. All most all of these cases of multiple meningiomas reported, showed multiple lesions at the time of operation or after a few years of the initial operation. Koech et al. (25) reported a case of 75-year-old female with two meningiomas.

Genetics is one of the important factors in development of multiple meningiomas (17). Some studies have reported that the deletion of the chromosome 22 (del22) in patients with type 2 neurofibromatosis and in up to 50% of solitary meningiomas is linked with the appearance of multiple meningiomas. Hormones also play an important role. There are a number of papers showing a higher frequency rate of meningiomas in women. Associated factor includes the action of progesterone on progesterone receptors found in eighty percent of meningiomas, leading to an increase in size during the luteal phase of the menstrual cycle and during pregnancy (18). The location of the tumours can be variable. There is a tendency towards unilateral hemispheric localization. The commonest locations were the supratentorial convexity and the para-sagittal falx. But multiple intracranial meningiomas occurring in the posterior fossa are very rare (19). Multiple intracranial meningiomas do not differ from the solitary types histologically (14); however, the simultaneous occurrence of different grades of malignancy in the nodules is observed in 1/3rd of multiple meningiomas (20). The commonest histological types reported in multiple meningiomas include psammomatous, fibroblastic, meningothelial, and transitional types (21).

The treatment and prognosis of multiple intracranial meningiomas do not differ from those of solitary benign tumours. Surgery being the treatment of choice for multiple meningiomas, depends on the following characteristics: symptomatic meningioma, asymptomatic meningioma which is greater than 3 cm in size and surgically accessible, and symptomatic expanding tumor (22, 23). Each tumour should be approached individually. therefore, mere presence of multiple tumours does not justify their removal. Since it is well documented that multiple meningiomas are almost always histologically benign, the prognosis is good and may be the same as for solitary meningiomas. Some studies showed no recurrence at follow-up evaluation (10, 14, 20).

Nakamura et al. (24) further highlight the natural history of asymptomatic meningiomas. The absolute annual growth rate ranges from 0.73 to 1.67 cc per year, and the tumour doubling time ranges from 1.19 to 6.81 years according to their study. This growth is minimal but varies with age and may thus be observed without surgical intervention unless specific symptoms develop. Asymptomatic small meningiomas should be followed up with MRI every 6 or 12 months if the patient is more than 65 years old. Surgery should be done mainly for an expansive symptomatic tumour with cerebral edema.

CONCLUSION

Multiple meningiomas is a rare entity by itself and incidence in males is extremely rare. Surgical treatment is the definitive line of mana gement. Timely intervention in those cases with large meningiomas adjacent vasogenic edema is of utmost importance. Small asymptomatic tumors may be left unoperated based on case to case scenario.

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