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Intracranial hemangiopericytoma in 26 years old female - A case report

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ABSTRACT

Hemangiopericytomas are uncommon central nervous system tumors of vascular origin. Surgery followed by radiotherapy is the mainstay of treatment. These tumors are known to recur locally and distant metastases are reported. Here, we are reporting a case of 26 years old female patient with clinical manifestation of headache and swelling over scalp. Radiological examination in the form of MRI suggested of hemangiopericytoma. The surgically removed tumor was sent to the histopathology department and final diagnosis was given as hemangiopericytoma.

Keywords: Hemangiopericytoma, Histopathology, Scalp swelling.

1. INTRODUCTION

Hemangiopericytoma is an uncommon tumor that was first described and named by Stout and Murrays in 1942, but it was not until 1949, when Stout reported 25 additional cases, that the tumor received widespread recognition (Stout et al., 1943; Stout et al., 1942). The hemangiopericytoma is thought to originate from the pericytes of Zimmermann (Stout et al., 1943; Stout et al., 1942; Conley et al., 1977; Nunnery et al., 1981; Enzinger et al., 1976) which are unique

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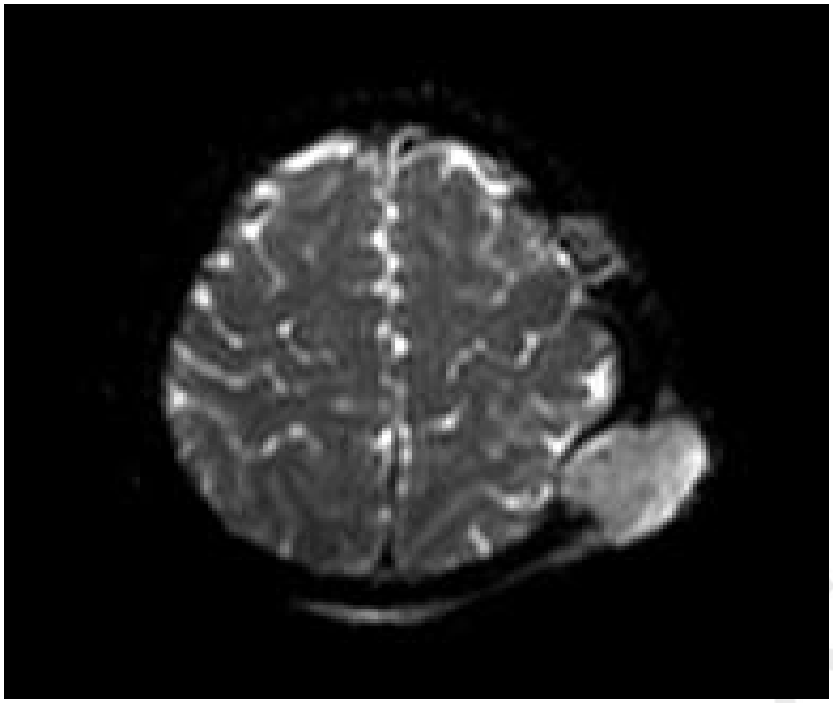

Figure 1
DWI showed multiple irregular hyper intense lesions

Figure 2
T2WI showed hyper intense lesions of hemangiopericytoma

cells found spiralling around the outside of blood capillaries and post-capillary venules. Central nervous system hemangiopericytomas are rare (Stout et al., 1942). Meningeal hemangiopericytomas constitute < 2.5 % of all meningeal tumors and < 1 % of all intracranial tumors (Guthrie et al., 1989; Jääskeläinen et al., 1985; Dufour et al., 2001). Surgery followed by radiotherapy is the mainstay of treatment. These tumors are known to recur locally and distant metastases are reported. As intracranial hemangiopericytomas are rare tumors and it is even rarer to find them in 26 years old, so here we are presenting a case where we are able to find and document the typical features of hemangiopericytoma.

2. CASE REPORT

A 26 years old female patient was apparently asymptomatic before 9 months and then she started to have swelling over scalp that progressively increased. She had also complaint of headache and dizziness for 8 months. Both general and neurological examinations were normal. Hematological and serological examinations were also within normal limits. In the last years, new techniques of neuro images and histopathological methods have been added to the diagnosis of cerebral mass lesions (Goyal et al., 2014). MRI of brain was performed. On DWI, multiple irregular hyper intense lesions were seen in the vault of the skull in left parietal bone, superior sagittal sinus and in the right side of the neck. On T2WI, lesions appeared hyper intense in nature with flow void areas within the lesions. Overall appearance suggested of multiple hemangiopericytoma involving skull, scalp and in right side of neck (Figures 1-3). The mass was excised and histopathological examination showed vascular spaces of variable sizes surrounded by plump to spindle cells, forming sheets of pericytes. The sheets of the cells showed presence of tiny capillaries in between. The cells showed little nuclear pleomorphism with few scattered mitotic figures 4-6. Overall histological findings were suggested of hemangiopericytoma.

3. DISCUSSION

Intracranial hemangiopericytomas are rare tumors arising from pericytes around capillaries or from mesenchymal cells. Silver staining techniques have demonstrated long processes extending from these cells and wrapping around the vessels. The exact function of the pericyte is not known, but it has been postulated that this cell has contractile powers and so controls the calibre of the vessels. Because the pericyte is found in the capillaries and venules of practically all types of tissue, it is not surprising that hemangiopericytomas have been reported as arising in many areas of the body. However,

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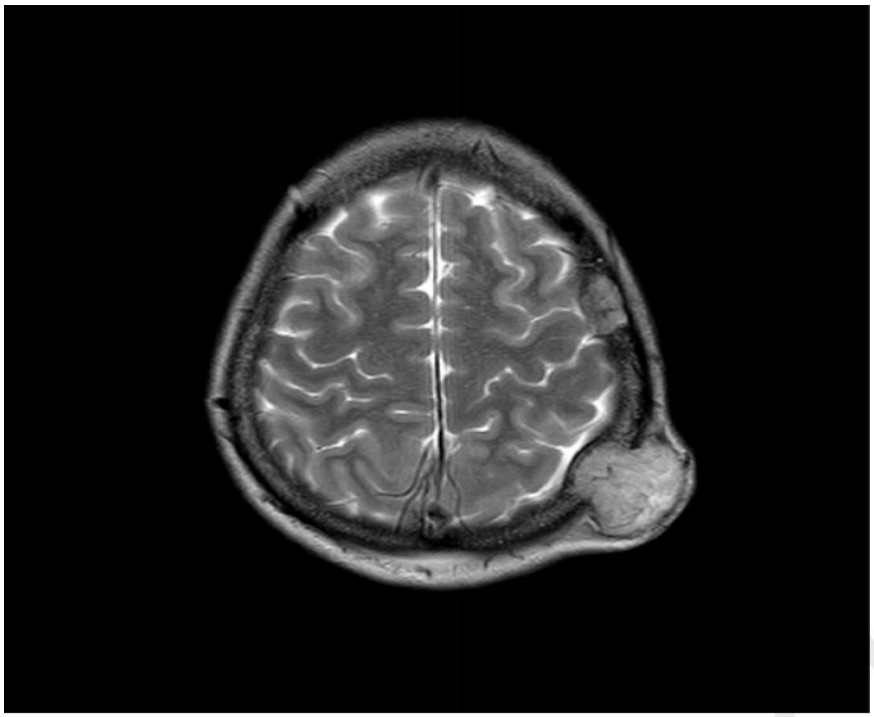

Figure 3

T2WI showed lesions with flow void areas

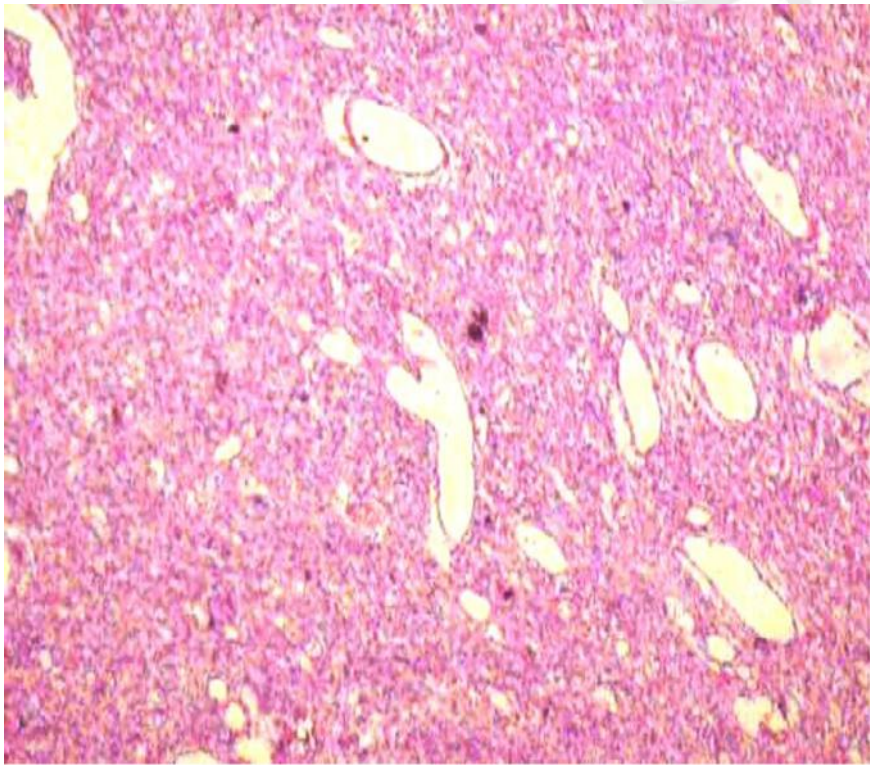

Figure 4

Presence of vascular spaces of variable sizes surrounded by sheets of pericytes. [H & E, 4 X]

hemangiopericytoma develops mainly in the lower extremities, retro peritoneum or pelvis and rarely occurs in the larynx, spleen, bone, meninges or thorax (Ciappetta et al., 1985; Schwartz et al., 1987; Sellke et al., 1991).

Meningeal hemangiopericytomas are largely tumors of adulthood particularly among men, and the average age at diagnosis ranges from 32 to 41 years in different series (Guthrie et al., 1989; Jääskeläinen et al., 1985; Kim et al., 2003; Mena et al., 1991; Herzog et al., 1995; Galanis et al., 1998; Pitkethly et al., 1970). But our patient was 26 years old female which is rather rare. Microscopically, the tumor mass consisted of tightly packed spindle shaped cells surrounding ramified thin-walled endothelium-lined vascular channels with characteristic 'antler' or 'staghorn' configuration. The neoplastic cells had round to oval to slightly angulated nuclei with clumped chromatin and a moderate amount of cytoplasm with ill-defined cell borders.

The differential diagnosis includes meningioma. We can differentiate hemangiopericytoma from the meningioma by presence of psammomatous calcospherites and tumor cells concentrically arrayed in tight whorl formations – both foreign to the hemangiopericytoma, as are the intra nuclear pseudo inclusions typical of meningothelial elements and their neoplastic derivatives. We can also differentiate these lesions from neoplasm of meningothelial lineage with use of an immunophenotyping that often includes extensive and strong labelling for CD99 and BCL2 but only focal, if any, EMA or claudin-1 expression (Mena et al., 1991; Hahn et al., 2006; Perry et al., 2005; Winek et al., 1989; Perry et al., 1997). The neoplastic cells of hemangiopericytoma are positive for vimentin, factor XIIIa, CD34, HLA-DR, and focally positive for desmin, actin, and myoglobin.

It is sometimes a locally aggressive, potentially malignant tumor that produces late local recurrences and distant extra neural metastases, mainly to bone and the lung and liver (Galanis et al., 1998; Mira et al., 1977; Staples et al., 1990). Meningeal hemangiopericytomas are aggressive tumors that tend to recur even after macroscopic total resection. Combination of wide local excision, radiotherapy and chemotherapy had been adopted in various institutions (Rathod et al., 2013). However surgery is the definitive modality of treatment, radiotherapy has been used to treat surgically unapproachable or recurrent tumors and to diminish local recurrence (Uemura et al., 1992). The local control, disease-free survival, and overall survival rates are much greater when patients receive radiotherapy (Guthrie et al., 1989; Brunori et al., 1997; Bastin et al., 1992; Staples et al., 1990; Alén et al., 2001).

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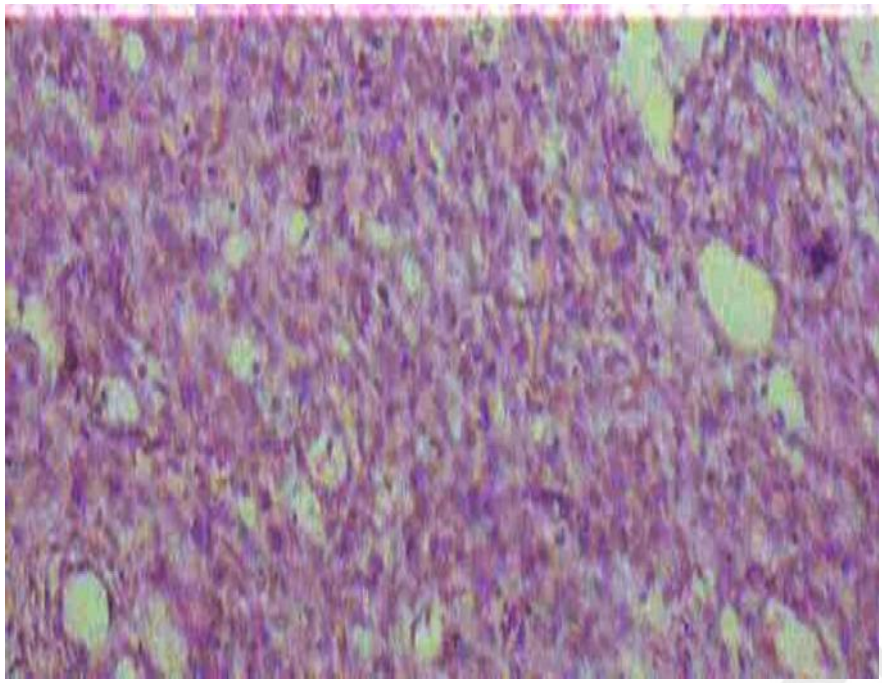

Figure 5

Plump to spindle cells forming sheets of pericytes. [H & E, 10 X]

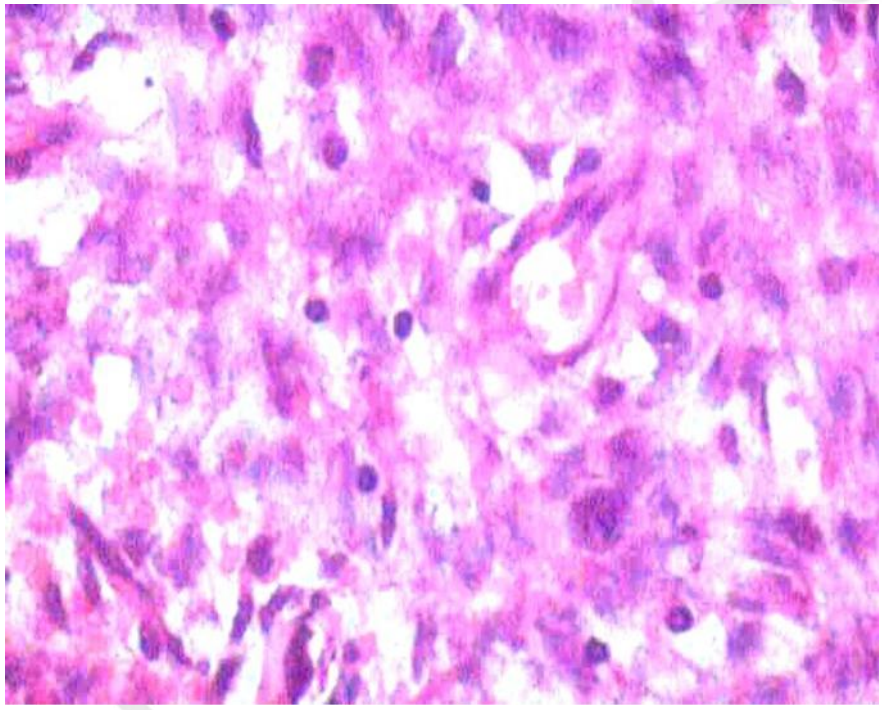

Figure 6

Presence of tiny capillaries in between the sheets of the cells. [H & E, 40 X]

Response to radiotherapy has been seen as long as 7 months after treatment. The 5 years survival rate as high as 71 % has also been reported (Spitz et al., 1998). Chemotherapy does not appear to have reputable role in the primary management of these tumors (Ecker et al., 2003). However in patients with disseminated distant metastasis palliative chemotherapy has been tried.

4. CONCLUSION

Hemangiopericytoma is uncommon central nervous system tumor of vascular origin. Sometimes they are locally aggressive and prone to recur also. Early and confirm diagnosis is must for better management of patient. Nowadays MRI and histopathology is the important diagnostic tool for the final diagnosis.

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