CASE REPORT

A Rare Case Report of Epithelioid Haemangioendothelioma of Ovary in Older Age Group

Anisa Barman[1], Sukla Naskar[2], Anadi Roy Chowdhury[3]

ABSTRACT

Epithelioid haemangioendothelioma is a very rare malignant vascular tumor with metastatic potential. It occurs between 30 – 50 years of age group. Most common sites are lung, liver, and bone. As a site of origin, ovary is the rarest. Few cases are reported till now. We received a specimen of total abdominal hysterectomy and bilateral oophorectomy in a case of 60 years woman, presented with left adnexal mass. Gross and microscopical features showed diagnosis of Epithelioid Haemangioendothelioma. The diagnosis is confirmed by immunohistochemistry study of CD34, Vimentin & Pancytokeratin.

KEY WORDS: Ovary, Epithelioid Haemangioendothelioma, CD34 positive.

Introduction

Epithelioid Haemangioendothelioma (EHE) is a rare malignant vascular tumor composed of epithelioid endothelial cells in a background of myxohyaline stroma. The prevalence rate is 1 in 1 million. It most commonly occurs in 30 to 50 years age group with a female predominance. Most common affected sites are lung, liver, and bone but any site or organ can be affected. Pathogenesis is predominantly WWTR1-CAMTA1 gene fusion & a small subset of tumors show YAP1-TFE3 fusion.

Case report

A 60 years old female presented in emergency department with pain in left iliac fossa region. After that she underwent for radiological investigations (Ultrasonography, CT scan) and CT scan report revealed this as a hyperechoic tumor of left ovary. After getting the radiology reports total abdominal hysterectomy and bilateral oophorectomy (TAH-BSO) was done on emergency basis. We received the specimen of TAH-BSO. Grossly the left ovary had a mass measuring 5 cm in maximum diameter. The cut section of the tumor showed haemorrhagic area. On histopathological examination section from the tumor showed lobulated vascular tumor comprising of epithelioid cells arranged in cords and nests in myxohyaline stroma. There were areas of anastomotic vascular channels. Tumor cells had moderate amount of cytoplasm with round nuclei and inconspicuous nucleoli. In some cells intracytoplasmic vacuoles were also seen. On immunohistochemistry study all the tumor cells showed CD34 membranous positivity & vimentin cytoplasmic positivity and PanCK negativity.

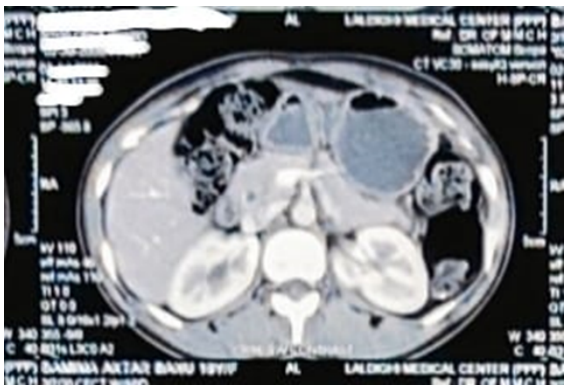

Figure 1: CT scan of ovarian mass

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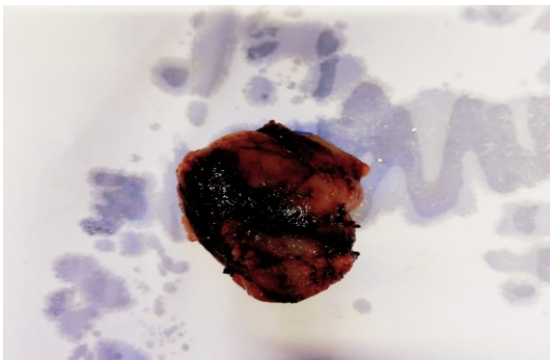

Figure 2: Gross picture of the mass

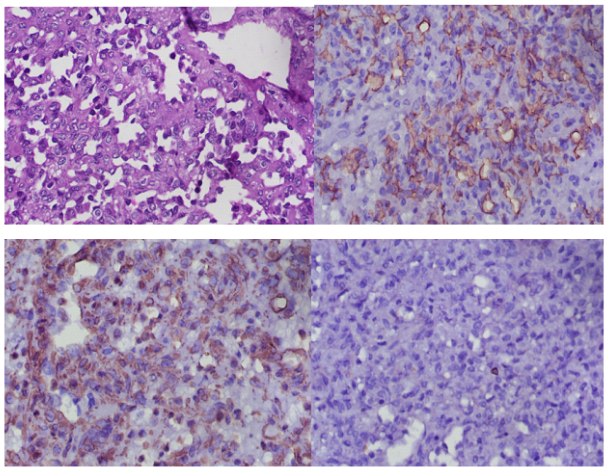

Figure 3: HP – Some cells have intracytoplasmic vacuoles (400X, H & E), CD34 – Membranous positive in tumor cells, Vimentin – Tumor cells show strong cytoplasmic positivity, PanCK – tumor cells are negative

Discussion

EHE is a very rare low grade malignant vascular tumor originating from endothelial or pre-endothelial vascular cells. It is an intermediate variant of the vascular tumor family. Benign end of this spectrum of disease is epithelioid hemangioma and the malignant end is epithelioid angiosarcoma. It can occur in both sexes however lung and liver lesions are more common in females, mostly in their second and third decades of life. There are no definite etiological factors but may be possibly associated with trauma, therapeutic radiation, and hormonal factors. Clinically it has the differential diagnosis of hemangioma, arteriovenous malformation, and hamartomas. Most cases are characterized by presence of WWTR1-CAMTA1 gene fusion, and a small subset of tumors is characterized by YAP1-TFE3 fusion. EHE was first described by Dail and Liebow as an aggressive bronchoalveolar carcinoma later recognized as pulmonary EHE. The involvement is more frequent in lung, liver, bone, and soft tissues but it can occur at any site or organ. In ovary it is very rare, till now 2 to 3 cases are reported in literature. Prognosis of EHE is variable. EHE having features of cellular atypia, mitotic activity (>1 mitotic figure per 10 high-power fields), necrosis and extensive spindling indicates an aggressive course and correlates with poor prognosis. Cases are always positive for at least one vascular endothelial marker. Definite diagnosis of our case was done by histopathological study and immunohistochemistry study by CD34, vimentin and PanCK[1–4].

Conclusion

We conclude the case as a diagnosis of Epithelioid Haemabgioendothelioma on basis of clinical findings of the patient, radiological investigations, and histopathological examination.

The diagnosis is confirmed by immunohistochemistry study by CD34, Vimentin and PanCK.

Limitations

As there are no facilities of molecular and cytogenetics study available in our institution, so we could not be able to do chromosomal analysis.

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