

Gastrointestinal Bleeding Caused By Duodenal Epithelioid Sarcoma: A Case Report

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Abstract

Case Report

Background: Epithelioid sarcoma is a rare malignancy that presents diagnostic difficulties. **Case:** A 22 years old female presented with epigastric pain, vomiting and haematemesis. Admitted for resuscitation and management, workup showed multiple duodenal and pancreatic hemangioma's underwent laprotomy end up with whipple's procedure, unfortunately she develop post op convulsion's, discovered to be due to brain metastasis, she passed away, histopathology came as epithelioid sarcoma. A review of different cases of similar condition is provided. **Conclusion:** The diagnosis of epithelioid sarcoma needs a high index of clinical suspicion and immunohistochemistry. Prognosis is determined by adequacy of the initial surgical procedure and early detection plays a major role and important predictor of recurrence.

Keywords: Gastrointestinal bleeding, epithelioid sarcoma, Classical type, proximal type, immunohistochemistry.

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INTRODUCTION

Epithelioid sarcoma is a rare soft tissue tumor with high susceptibility for local recurrence and distant metastasis. Constitute less than (1%) of all soft tissue sarcomas. commonly affects the distal extremities.

Lesions located or extending into the dermis often ulcerate through the skin and may simulate an ulcerating squamous cell carcinoma, especially because of the pronounced epithelioid appearance and eosinophilia of tumor cells [1-3].

Small bowel tumours are extremely rare, accounting for 1-2% of gastrointestinal tumours. Epithelioid variant is classified under tumours of uncertain differentiation in the WHO classification of tumours [4-6].

CASE PRESENTATION

A – 22- year Sudanese women presented with 4 weeks history of epigastric pain, burning in nature, localized aggravated with spicy food, associated with severe nausea, anorexia, retching and non- bilious vomiting. She was seen gastroenterologist plan for OGD, done on (12-10-2017) diagnosed as duodenal

ulcer and plan for PPI and review in 4 weeks interval. On (25-10-2017) she presented to algawda hospital with 5 day history of haematemesis contains food particles and clots; it was minimum then increase in amount and frequency. Associated with malena, dull ache central abdominal pain, retching, generalized malaise and fatigability.

Upon her admission she was very ill, very pale not jaundiced or distressed and fully conscious. BP= 100/60 mmHg, HR= 112bpm.

Abdominal examination: soft non distended abdomen with eigastric tenderness but no guarding, rigidity, rebound tenderness and no palpable mass. LNs weren't palpable. DRE: empty rectum and no palpable mass, blood or malena. Other systems were normal.

Her investigations showed hemoglobin of 7.4 g/dl (normochromic normocytic). Plan for urgent admission, transfusion and Re OGD, done on (26-10-2017) showed normal oesophagous and stomach, duodenum showed massive clot or tumor, difficult to identify due to unwashable blood, lesion wasn't amenable to clipping or heating, so biopsy taken and adrenaline injection applied(conclude as massive

duodenal clot/tumor). Recommend to perform CT abdomen which was done prior to OGD on (24-10-2017) showed (Multiple hepatic centripetally enhancing benign featuring focal lesion of hemangiomas, multiple bilateral benign featuring renal lesions either of angiomyomas or angiomyolipomas with scanty lipid content, pancreatic head benign featuring lesion mostly of hemangioma and two right sided pulmonary lesions mostly of hemangioblastomas. Picture may be in the course of phacomatosis either caused by tuberous sclerosis, von Hippel landau disease or neurofibromatosis type1).

Her condition continues to deteriorate with frequent episodes of hematemesis, malena and abdominal pain. Her hemoglobin level continues to drop and repeated blood transfusion were given, The patient was subsequently put up for exploratory laprotomy (28-10-2017) and the finding was two nodules at 2nd part of the duodenum no obvious other lumps, peritoneal seeding, liver secondary's and ascites, so Whipple's procedure was done for her successfully (figure 1). Smooth recovery, transferred immediately to ITU for monitoring. Her condition showed remarkable improvement in the subsequent three days, on the fourth's day, she start to take sips of fluids.



Fig-1: Show the specimen after resection

At same day she start to develop sever bilateral headache associated with frequent bilious vomiting alternating with black color vomitus and her level of consciousness start to deteriorate, there were no weakness or parathesia. Her abdomen was soft, no tenderness of distension and positive bowel sounds and drains were empty too. CT abdomen and brain done on (5-11-2017) showed (Enlarged liver with numerous deferent size hepatic scattered throughout the liver represented hemorrhagic metastasis, similar lesions are

seen at the right lung, similar mass is seen at the greater curvature of the stomach and enhancing mass is seen at each kidney. CT Brain (there are four hyperdense left cerebral masses with significant surrounding edema and midline shift to the left, representing metastasis.

In Conclusion features of a hemorrhagic metastatic disease to the liver, lung, kidneys, stomach and brain. And moderate hemorrhagic ascites. Unfortunately her condition continue to deteriorate with continues severe headache, decrease in level of consciousness and she arrested and passed away on (6-11-2017).



Fig-2: Show brain metastasis

Histological

Macroscopic description: A: segment of small intestine 12cm with central open part containing a black nodule 1c. B: piece of gray tissue.

Microscopic description: A section show haemorrhagic submucosal tumor formed of large cells, most with clear cytoplasm and big vesicular nuclei having prominent nucleoli. Some nuclei are larger and pleomorphic. The tumor is not infiltrating the muscle coat. Upper and distal margins of excision are clear. There are wide areas of haemorrhages and necrosis.

B: shows pancreatic tissue, no lymphoid tissue is seen

Diagnosis: Epithelioid sarcoma

DISCUSSION

Epithelioid sarcoma is a rare, high grade, soft tissue tumor with high propensity for local recurrence, regional lymph node involvement and distant metastasis. Types of epithelioid sarcoma are classical type, proximal type and distal type. The proximal-type ES is more aggressive than its classical counterpart. It

accounts for only one third of the total share of epithelioid sarcoma [7].

The classical-type most commonly affects the distal extremities. It never affects the trunk, genitals and head and neck regions. This type has a distinct nodular arrangement of eosinophilic spindle and epithelioid cells and has a tendency to undergo central degeneration and necrosis [8].

The clinical presentation of epithelioid sarcoma is usually nonspecific and that made its management is challenging. Epithelioid sarcoma may disseminate via the sub dermal lymphatic vessels or the blood stream, as seen in our patient where the lesion was scattered in many organs [9].

Diagnosis of epithelioid sarcoma is made via imaging using CT scan or magnetic resonance imaging (MRI) scan often to look for the cause of symptoms, to find tumors or to stage the disease. PET scan is also used for its ability to identify unknown primary and unusual metastatic sites. CT Angiogram of the mesentery was used in some cases to detect the probable cause of the gastrointestinal bleed as part of GIT bleeding workup [4].

Chbani *et al.* described that valentine reactivity is present in almost all cases, while pan keratin AE1/AE3 and epithelial membrane antigen were positive in 96% and 98% of the cases respectively [10]. Immunohistochemistry remains the golden test to confirm the diagnosis [1].

The prognosis of epithelioid sarcoma is very poor due to its aggressive nature, and late presentation of patients with a local recurrence rate of 85 percent and distant metastatic rate of 30 percent, it's known to propagate along tendon and nerve sheaths and fascial planes. Palliative surgical resection is the main surgical modality, since complete resection is often not possible in many cases due to the aggressive infiltrative nature of the tumors [5, 9]. Nevertheless; large trials is recommended to improve the management's guidelines of this disease.

CONCLUSION

Epithelioid sarcoma is difficult to diagnose and a high index of suspicion is needed. Immunohistochemistry is an extremely useful tool to confirm the diagnosis. Prognosis is determined by adequacy of the initial surgical procedure and early detection plays a major role and important predictor of recurrence.

REFERENCES

1. Dhaval Jetly, Annapoorani Varadarajan, Kinnari S Naik, Priya Patel. Epithelioid sarcoma: A case series of three different presentations. *Case Reports in Clinical Pathology*. 2017; (4): 3.
2. Karagkounis G, Argyrakos T, Charkiolakis G. A case of distal epithelioid sarcoma of the thumb expressing podoplanin, TLE1 and Ca 125. *Case Rep Pathol*; 2013.
3. Armah HB, Parwani AV. Epithelioid sarcoma. *Arch Pathol Lab Med*. 133(5): 814-9.
4. Lee Fei Yee MD, Lim Limi, MS, Tikfu Gee, MS. Gastrointestinal bleeding caused by epithelioid sarcoma: A case Report. *Med J Malaysia*. 2016; (71): 4.
5. Lin CF, DeFrias D, Lin X. Epithelioid angiosarcoma: a neoplasm with potential diagnostic challenges. *Diagn Cytopathol*, 2010; 38(2): 154-8.
6. Fletcher C. Guillou L, Kaneko Y. Epithelioid sarcoma. In: Fletcher C. WHO. Classification: Tumors of Bone and Soft Tissue. Geneva, Switzerland: World Health Organization; 2002.
7. Bhama BA, Dhir RS, Pandit AA. Epithelioid sarcoma of the upper extremity (a case report). *J Postgrad Med*. 1987; 33(3): 140.
8. Suárez-Zamora DA, Barrera-Herrera LE, Rodríguez-Urrego PA. Proximal-Type Epithelioid Sarcoma: Report of an Unusual Case in the Uterine Cervix. *Int J Surg Pathol*. 2017 Feb 1.
9. Enzinger FM. Epithelioid sarcoma: a sarcoma simulating a granuloma or carcinoma. *Cancer*. 1970; 26: 1029-41.
10. Chbani L, Guillou L, Terrier P. Epithelioid sarcoma: A clinicopathologic and immunohistochemical analysis of 106 cases from the French Sarcoma Group. *Am J Clin Pathol*. 2009; 13: 222-227.