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Undifferentiated High Grade Spindle Cell Sarcoma of the Ovary Arising in the Background of Teratoma

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Case Presentation

A 60 years old postmenopausal woman presented with abdominal pain and vomiting. CT abdomen pelvis showed pelvic mass with suspicious of ovarian teratoma. MRI study of pelvis pre and post contrast revealed a large lobulated mixed signal intensity mass lesion of 15.5x10.9x15.7 cm of size in the pelvis extending into the abdomen up to level of T4 vertebral body. Mass had heterogeneous signal intensity solid and cystic components within it. Solid component was isointense to muscle on T1 weighted images. Cystic component measured 5.8x6.1 cm. No definite contrast enhancement seen. Superiorly lesion reaches up to the under surface of mesentery. Ovaries were not visualized separately suggestive of complex neoplastic mass arising from ovaries. Case of obstructive incarcerated hernia, with pelvic mass.

Emergency surgery done finding incarcerated omentum in umbilical hernia, resection of the omentum, dilatation of the transverse and descending colon due to obstruction of sigmoid by a complex solid and cystic mass arising from the left ovary, decision to do debulking total abdominal hysterectomy with resection of the obstructed part of the sigmoid, end to end anastomosis done with endo GIA, cytoreductive surgery to all macroscopic lesion in the pelvic peritoneum.

Macroscopically, left ovary was markedly enlarged measuring 17x14x10 cm with multilobulaiton. (Figure 1) Cut surface was predominantly solid with gray white lobulated surface and pale yellow areas. Necrotic areas with cystic changes and calcification is noted. Large cystic area measuring 7x7x5 cm and attached nodule measuring 6 cm showing marked myxoid changes. Attached colon was 11 cm in length. (Figure 2).

Figure 1. Gross image of left Ovary: Note the predominantly solid and cystic areas of the tumor

Figure 2. Cut section of ovary: Cystic areas with mural nodules. Colon is adherent to the tumor

Histological section showed neoplasm composed of pleomorphic spindle cells forming fascicles and sheets. The spindle cells show marked pleomorphism with epithelioid morphology in areas with round vesicular nucleus with prominent nucleoli and moderate eosinophilic cytoplasm. (Figure. 3) Frequent mitosis and patchy are of necrosis was seen. (Figure. 4) Myxoid area sections showed necrotic tumor with hyalinization. The neoplasm is seen infiltrate to attached sigmoid colon. (Figure 5) Infiltration was up to muscularis propria. Tumor had pseudo capsule all around and capsule surface shows foreign body giant cell reaction.

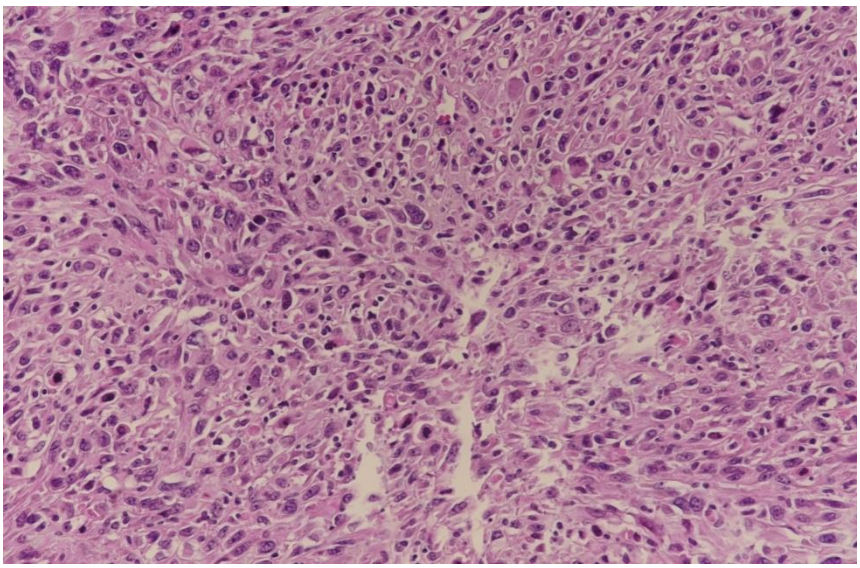

Figure 3. High power showing marked pleomorphism of the spindle cells and frequent mitosis.

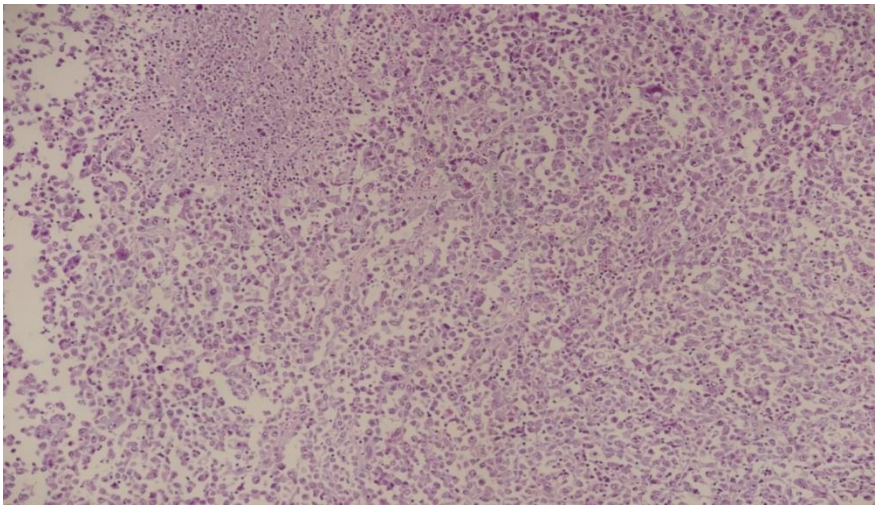

Figure 4. Tumor shows extensive areas of necrosis.

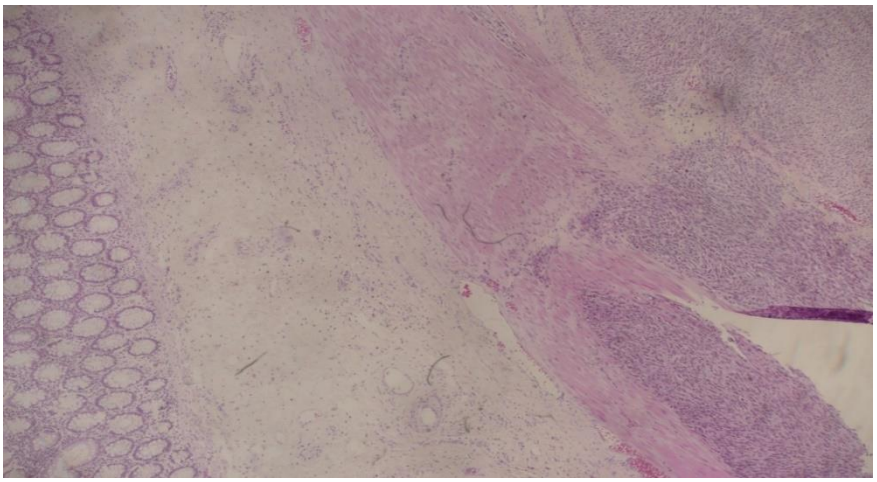

Figure 5. Tumor cells are seen to infiltrate the muscle layer and reaches submucosa of the colon

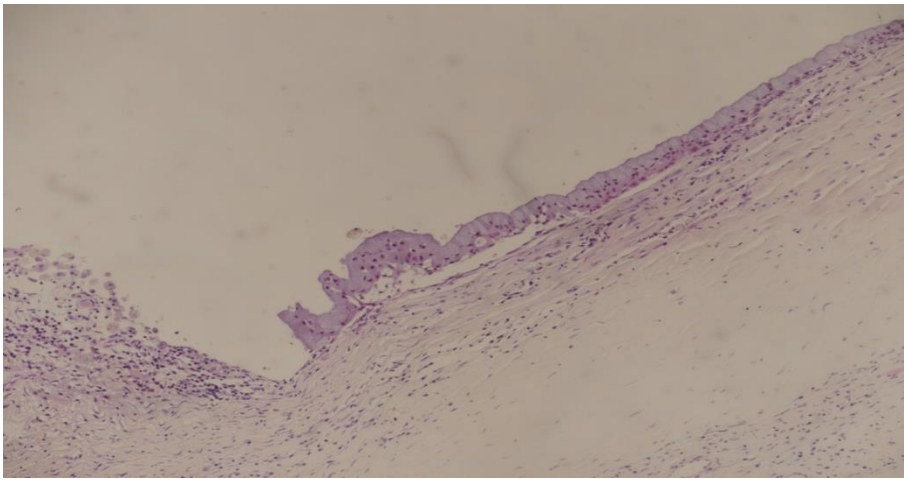

Figure 6. Shows mucinous lining of the cystic areas with ulceration of the lining

Immunohistochemically, tumor cells were negative for Pan Cytokeratin (clone:AE1/AE3), CK7 (clone: OV-TL 12/30) and CK 20 (clone: Ks20.H) suggested negative for epithelial differentiation. CD117 (clone: polyclonal) and CD34 were negative and ruled out gastrointestinal stromal tumor as tumor is infiltrating sigmoid colon. Melan A and HMB 45 were negative ruling out melanoma. Tumor cells were also negative for Desmin, CD31, CD34, S100 and SMA. Ki-67 was 80-90%. Further immunohistochemistry was negative for MDM2 (for liposarcoma), Inhibin (for granulosa/sertoli cell tumor), STAT6 (for solitary fibrous tumor), Myogenin (for Rhabdomyosarcoma), Calretinin (for mesothelioma) and CAM5.2 (Myofibroblast and smooth muscle tumor). Histologic features and immunoprofile were suggested of undifferentiated high grade pleomorphic spindle cell sarcoma of left ovary arising in the background of teratoma. There was no specific differentiation towards skeletal muscle, smooth muscle or vascular structure. Peritoneal mass biopsy showed similar pleomorphic spindle tumor cells as in left ovary. Some area showed necrotic tumor cells with secondary hyalinization.

Discussion

Germ cell tumors account for approximately 30% of all ovarian tumors. According to WHO, malignancy arising within a mature cystic teratoma is a rare complication seen in <1-2% cases. The most common malignant change in cystic teratoma is Squamous cell carcinoma (75%), followed by carcinoid tumor, adenocarcinoma and melanomas. Sarcomas account for 8% malignancies in dermoid cysts. **[**3**]**

The frequency of malignant change increases with increasing age, rising to 19% in women after menopause. **[**4**]**

Study by Malagon et al., the most common sarcomatous component to arise in germ cell tumors was embryonal rhabdomyosarcoma (29/46) followed by angiosarcoma (6/46), leiomyosarcoma (4/46) and undifferentiated sarcoma (3/46). **[**5**]**

Patient with sarcoma often present late and can grow large without producing symptoms. We report a case of poorly differentiated spindle cell sarcoma in background of teratoma which is very rare. Case presented with nonspecific abdominal pain and vomiting. Patients with ovarian sarcoma had a mean age of (54.3 ± 10.3) years, and 16 of them were postmenopausal. **[**6**]**

In the differential diagnosis of these spindle-shaped lesions, we should also consider the possibility of gastrointestinal stromal tumors involving the ovary; in these cases, immune stain for CD117 or molecular studies, evaluating c-kit mutations, would be essential. [7,8]

Other processes that eventually affect the ovary are endometrial stromal sarcomas which must be taken into account, especially those with a spindle shaped cell pattern. These cases will need immunohistochemistry with CD10 to demonstrate their stromal nature. [9]

some tumors with occasional spindle cell or sarcomatoid pattern can involve the ovary such as dendritic cell neoplasms and melanomas, where immunohistochemical stains will be also necessary. [10,11] Finally, ovarian metastases of sarcomas originating in other locations should also be ruled out.

Conclusion

We report a very rare occurrence of sarcomatous malignant transformation in surgically resected ovarian mass suspected to be mature cystic teratoma and presenting as acute abdomen. Neither imaging studies nor tumor marker levels can accurately diagnose such a malignant transformation pre-operatively. Hence all the dermoid cysts should be examined histologically by giving multiple sections with particular focus on solid areas along with the aid of immunohistochemistry in case of poorly differentiated tumors. This will result in more such cases being identified and thus treated at an early stage.

Post operatively, Patient is doing well. She was discharge from the hospital at day 10 post-surgery, without any complication. After final result of histopathological examination, MDT decision is to start chemotherapy.

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