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CASE REPORT

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Primary ovarian angiosarcoma: two case reports

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Abstract

Background: Primary angiosarcoma of the ovary is extremely rare, and its pathologic diagnosis is challenging due to its non-specific clinical symptoms, and morphologically. **Cases**: In this report, we presented the clinicopathological characteristics of 2 cases of primary ovary angiosarcoma. In case 1, a 31-year-old patient with abdominal pain for 1 month and a cystic-solid mass with a diameter of about 15 cm in the right uterine adnexa was described. After surgery, a right ovarian angiosarcoma of IA clinical stage was confirmed. The patient received 6 rounds of chemotherapy after surgery and was now alive 9 years with no evidence of recurrence. Case 2 was a 32-year-old patient presenting with abdominal pain and a cystic-solid mass in the left ovary with a diameter of 14 cm was detected. Pathological diagnosis confirmed left ovarian angiosarcoma of Federation International of Gynecology and Obstetrics (FIGO) stage IIIB. After 2 rounds of chemotherapy, the tumour relapsed and the patient died 7 months after the initial diagnosis. The morphology of the tumour was heterogenous in different areas, from well-differentiated areas of angioma-like benign morphology, to poorly differentiated spindle cell areas. Careful observation could find the formation of microcystic or fissure-like structures formed by blood vessels. Immunohistochemically, the tumour cells were diffusely positive for CD31, Fli-1, ERG, and factor VIII (FVIII). CD34 Showed diffusely positivity in case 1, and focal positivity in case 2. Some tumour cells expressed vascular endothelial growth factor (VEGF) and D2-40. Expression of epithelial markers along with SALL4, α-inhibin, calretinin, S-100 and desmin were all negative. The Ki-67 indexes of the 2 patients were 80% and 60%, respectively. **Conclusions**: The accurate diagnosis of primary ovarian angiosarcoma requires careful histomorphologic examination and reasonable immunohistochemistry staining.

Keywords

Ovary; Angiosarcoma; Immunohistochemistry; Pathology

1. Introduction

Angiosarcoma is a rare mesenchymal tumour and accounts for approximately 2% of all soft tissue sarcomas [1]. It occurs mainly in the skin and superficial soft tissues of the head and neck in elderly patients but can also occur in the heart, liver, spleen, lung, gastrointestinal tract, and female genital tract [2]. Primary ovarian angiosarcoma is extremely rare. The diagnosis of this tumour is challenging because of its lack of specific clinical symptoms, complex and varied morphology and morphological similarity to other malignant tumours of the ovary. It progresses rapidly, and patients have poor prognoses and outcomes. In our report, we present the clinical manifestations, pathological characteristics, and clinical prognosis of 2 cases of primary ovarian angiosarcoma to better understand this type of rare malignant tumour.

2. Materials and methods

The two cases were retrieved from West China Second University Hospital, Sichuan University. The available clinicopathological information, including age, clinical presentation, tumour size, and follow-up data, was collected. The tissue blocks were submitted in whole or in part for pathological examination. Haematoxylin and eosin (HE) stained slides were independently evaluated by two pathologists. Immunohistochemical staining was performed with the following antibodies (Table 1) on a Roche Ventana staining system.

3. Case report

Case 1 is a 31-year-old female (gravida 2 para 1 (G2P1)) who sought treatment due to “lower abdominal pain for 1 month”. Vaginal ultrasound suggested a cystic-solid mass in the right uterine adnexa. The serum cancer antigen 125 (CA125) concentration was 69.0 U/mL (normal value <35 U/mL), and the cancer antigen 19-9 (CA 19-9) concentration was 47.1

TABLE 1. Antibody information.

	Full name	Source	Clone	Dilution
CD34	Cluster of differentiation 34	DAKO	QBEnd/10	1:100
CD31	Cluster of differentiation 31	Roche	MXO32	1:200
FLI-1	Friend leukaemia integration-1	Leica	G146-22	Undiluted
ERG	ETS related gene	Zhongshan	UMAB78	1:400
D2-40	D2-40	Roche	D2-40	1:50
CK	Cytokeratin	DAKO	AE1/AE3	1:400
EMA	Epithelial membrane antigen	DAKO	E29	1:100
CK8	Cytokeratin8	Roche	TSI	1:150
CK7	Cytokeratin7	Leica	OV-TL12/30	1:200
CK18	Cytokeratin18	Roche	MX035	1:150
SALL4	Spalt like transcription factor 4	DAKO	6E3	Undiluted
α-Inhibin	α-Inhibin	DAKO	MXO98	1:50
Calretinin	Calretinin	DAKO	MX027	1:50
Desmin	Desmin	DAKO	MX046	1:100
S-100	S-100	Roche	4C4.9	1:200
AFP	Alpha fetoprotein	DAKO	EP209	Undiluted
Ki-67	Ki-67	DAKO	MXR002	1:300

U/mL (normal value <30.9 U/mL). The patient underwent laparotomy with right adnexectomy, and the right ovary was found to be occupied by a cystic-solid mass with a diameter of approximately 15 cm. Intraoperative frozen section diagnosis suggested a malignant tumour favouring sarcoma, and yolk sac tumour needed to be excluded. There were no obvious abnormalities in the uterus and left adnexa. Subsequently, a fertility sparing surgery including greater omentectomy and pelvic lymph node dissection was performed. Postoperative pathology and computed tomography (CT) confirmed that the tumour was confined to the ovary, and there was no evidence of extraovarian involvement. The capsule of the tumour was intact after complete resection of the right adnexa. No tumour cells were found in the ascites. Further hysterectomy and contralateral oophorectomy were declined due to the patient's reproductive needs. Gross examination showed a reddish-brown mass on the right ovary with a volume of 15 cm × 10.5 cm × 7.5 cm with extensive bleeding and necrosis. The cutting surface was solid with a grey-brown colour and a soft texture, and some areas were microcystic or spongy. The pathological diagnosis was International Federation of Gynecology and Obstetrics (FIGO) stage IA right ovarian angiosarcoma. Three months after surgery, no abdominal space occupation and no abnormalities in the left adnexa and uterus were identified by CT scanning. The patient received 6 rounds of Adriamycin + Ifosfamide chemotherapy after surgery and remains alive 9 years after surgery, with a yearly CT scan and no evidence of recurrence.

Case 2 is a 32-year-old female (G0P0) who sought treatment due to "lower abdominal pain for more than 3 months". Colour ultrasound suggested an irregular mass with abundant blood flow signals and a size of approximately 8.0 cm × 7.9 cm × 5.9 cm in the left posterior uterus (Fig. 1A). Pelvic examination showed a 2.1 cm collection of fluid appearing as a sonolucent area. Laboratory examination showed normal levels of tumour biomarkers, including CA125, CA 19-9, alpha-fetoprotein (AFP), and human chorionic gonadotropin (HCG). The patient initially underwent laparoscopic exploration, which revealed a solid mass of approximately 10 cm in the left ovary that was tightly adhered to the sigmoid colon, rectum, and uterus. A biopsy tissue was taken from the tumour and intraoperative frozen section diagnosis suggested a malignant mesenchymal tumour. Based on this diagnosis and in order to avoid rupture of the mass and tumour spillage, the surgeon then decided to perform laparotomy for subtotal hysterectomy and bilateral adnexectomy with omentectomy, bilateral pelvic and para-aortic lymphadenectomy and appendectomy. Adhesion between the anterior wall of the rectum and the posterior wall of the uterus was also observed, and a 2 cm × 1 cm area of rough texture on the serous surface of the anterior wall of the rectum was completely removed to ensure radical (R0) resection. Gross examination showed a brown cystic-solid mass with a volume of 14 cm × 10 cm × 2.5 cm in the left ovary (Fig. 1B). The inner wall had a rough texture, and many blood clots were observed. Pathological diagnosis showed left ovarian angiosarcoma involving the omentum, uterine serosal layer and serous surface of the left anterior wall of the rectum. The tumour was classified as FIGO stage IIIB.

After surgery, the patient received 2 rounds of Adriamycin + Ifosfamide chemotherapy. Three months later, the CT results suggested tumour recurrence and spread in the abdominal cavity, as evidenced by multiple low-density shadows indicating soft tissue in the pelvic and bilateral rectus abdominis muscles and local nodular shadows in the anterior peritoneum. The patient did not receive further adjuvant therapy, and she died 7 months after the initial diagnosis.

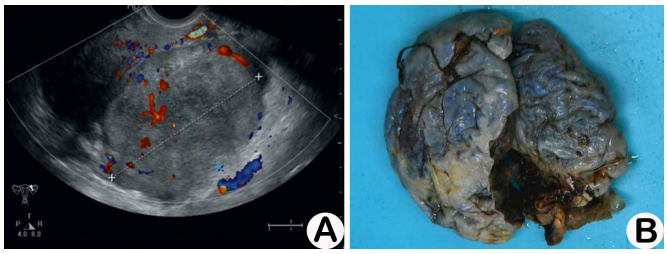

FIGURE 1. B-ultrasound images and gross picture of tumour. (A) Colour ultrasound of case 2 suggested a mass with slightly weak, uneven echo and a volume of approximately 8.0 cm × 7.9 cm × 5.9 cm in the left posterior uterus. (B) Gross examination showed a brown cystic-solid mass in the left ovary with a volume of 14 cm × 10 cm × 2.5 cm.

Microscopic examination indicated that all tumour tissues in these 2 cases were contained within the ovarian parenchyma and exhibited infiltrative growth. The tumours exhibited a heterogeneous morphology. In case 1, the well-differentiated areas contained many vascular lumen structures of different sizes. Some of these lumen structures were dilated and filled with red blood cells (Fig. 2A). The lining of the cystic wall contained mildly atypical tumour cells. Nuclei were flat or slightly enlarged and could also stack to generate papillary protrusions or a hobnail shape (Fig. 2B). Some areas were poorly differentiated, but formation of vascular lumen structures was observed even in these areas. Scattered multinucleated giant tumour cells could also be seen (Fig. 2C). A few areas were composed of foam-like cells with vacuolated cytoplasm (Fig. 2D). Immunohistochemically, the tumour cells were diffusely positive for CD31 and CD34 (Fig. 2E,F), ERG (Fig. 2G), FLI-1 (Fig. 2H) and factor VIII (FVIII). In case 2, the left ovary was almost completely replaced by tumour tissue, and residual ovarian tissue was observed only in some local areas (Fig. 3A). The tumour was poorly differentiated, and only a few regions contained clearly differentiated vascular structures (Fig. 3B). Most regions consisted of spindle-shaped tumour cells that formed irregular cystic tubular or fissure-like structures and cross-linked and communicated with one another (Fig. 3C). In addition, round or polygonal cells with a patchy distribution were observed within the tumour (Fig. 3D). Necrosis and extensive bleeding were also observed. Tumour cell atypia was obvious, and mitotic activity was high, with counts of approximately 5-10 mitoses/10 high-power fields. Immunohistochemically, the tumour cells were diffusely positive for CD31 (Fig. 3E), CD34 showed focal positivity (Fig. 3F). In addition, tumour cells also exhibit diffuse expression of ERG (Fig. 3G), FLI-1 (Fig. 3H) and factor VIII (FVIII).

In the two cases, some tumour cells expressed vascular endothelial growth factor (VEGF) and D2-40. Epithelial markers, including CK, EMA, CK7, and CK8/18, all exhibited negative expression. The immunostaining results for SALL4, AFP, α-inhibin, calretinin, S-100, and desmin were negative. The Ki-67 indexes in case 1 and case 2 were 80% and 60%, respectively.

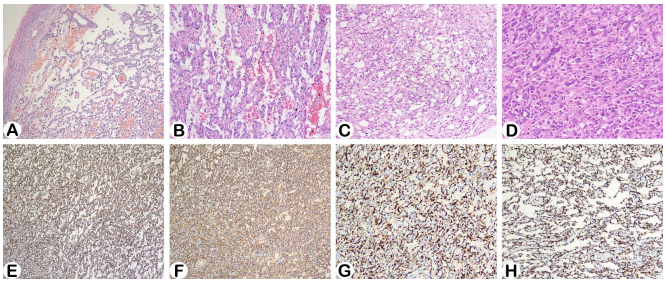

FIGURE 2. Histology and immunohistochemical staining pictures of the tumour of case 1. (A) The well-differentiated areas contained many vascular lumen structures of different sizes (haematoxylin-eosin (HE) staining, 40×). (B) Some of these lumen structures were dilated and filled with red blood cells. The lining of the cystic wall contained mildly atypical tumour cells (HE staining, 100×). (C) A few areas were composed of foam-like cells with vacuolated cytoplasm (HE staining, 100×). (D) Some areas were poorly differentiated, with scattered multinucleated giant tumour cells (HE staining, 200×). Tumour cells was diffusely positive for CD31 (E), CD34 (F), ERG (G), FLI-1 (H) (IHC staining, 100×).

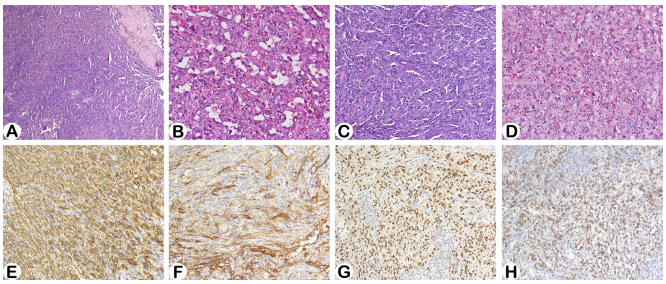

FIGURE 3. Histology and immunohistochemical staining pictures of the tumour of case 2. (A) The ovary was almost completely replaced by tumour tissues, and residual ovarian leukosis was only observed in some local areas (haematoxylin-eosin (HE) staining, 40×). (B) In some regions the tumour showed clearly differentiated vascular structures (HE staining, 100×). (C) Most regions consisted of spindle shaped tumour cells which formed fissure-like structures (HE staining, 100×). (D) Round or polygonal cells with a patchy distribution were observed in some local area side the tumours (HE staining, 200×). Tumour cell showed diffusely positivity for CD31 (E), focal positivity for CD34 (F), ERG (G) and FLI-1 (H).

4. Discussion

Angiosarcoma rarely occurs in the female reproductive tract; however, it can occur in the uterus, vagina, vulva, cervix, and ovary. Primary ovarian angiosarcoma is extremely rare. To date, only 32 cases of primary ovarian angiosarcoma have been reported in the literature [3–25]. Angiosarcoma can occur at any age, and the age range reported in the literature is 11–81 years (mean age of 33 years). Most patients are women of reproductive or premenopausal age. The clinical symptoms are nonspecific, and the major presentation is abdominal pain and distension. In most cases, the mass is unilateral but can rarely be bilateral [16, 20]. Masses are slightly more common in the right ovary than in the left ovary [25]. The tumours generally have a large volume, the maximum diameter can reach 30 cm, and the mean diameter is approximately 14 cm.

Histologically, primary ovarian angiosarcoma can be pure angiosarcoma, although some angiosarcomas are associated with other tumours, such as mature cystic teratoma and mucinous or serous tumours [5, 20, 21]. The morphology of the tumour can vary greatly between patients and in different areas in the same patient. The tumour can have an angioma-like benign morphology, a cystic tubular structure with fissure or maze-like mutual communication, or poorly differentiated solid areas containing spindle-shaped cells. The poorly differentiated areas may be similar to those observed in spindle cell sarcoma or spindle cell carcinoma. Sometimes it is difficult to immediately consider these tumours as vascular tumours. However, careful observation can reveal the presence of microcystic or fissure-like structures formed by blood vessels.

By immunohistochemistry, the tumour cells can be found to express vascular endothelial markers, including CD31, CD34, FVIII, ERG, and FLI-1. Some tumours can also express D2-40 and VEGFR3, which indicate lymphatic endothelial differentiation. The CD31-positive rate in angiosarcoma is 90%, and the CD34-positive rate is 50%–60% [24]. CD31 has high sensitivity and specificity, especially in poorly differentiated angiosarcoma. However, notably, histiocytes, plasma cells, and multinucleated giant cells can also express CD31. ERG expression has high sensitivity and specificity in angiosarcoma. FLI-1 has excellent sensitivity, but its specificity is slightly lower. However, both ERG and FLI-1 can also be expressed in other tumours of nonvascular origin [26, 27]. Therefore, the combination of CD34, CD31, ERG and FLI-1 is more helpful for accurate diagnosis than any of these markers alone.

Regarding differential diagnosis, highly differentiated angiosarcoma is histologically similar to ovarian angioma, which is rare and can occur at any age, with a tumour diameter ranging from several millimetres to several centimetres. Histologically, these tumours are usually spongy and contain flat lining endothelial cells without atypia and mitosis. An important indicator for distinguishing angioma from well-differentiated angiosarcoma is whether the tumour has peripheral tissue infiltration. In addition, juvenile haemangioendothelioma should also be excluded. One case of juvenile ovarian haemangioendothelioma in a newborn has been reported [28]. The tumour contained vascular lumens with varying sizes, and the cytoplasm of the vascular endothelial cells was eosinophilic. The tumour cells showed vesicular nuclei, and small nucleoli and mitosis were occasionally observed, with no atypia. Poorly differentiated angiosarcoma needs to be differentiated from sarcomatoid carcinoma, malignant melanoma, and leiomyosarcoma. Morphologically, the vascular lumen structures containing red blood cells in angiosarcomas should be carefully identified. Immunohistochemistry can also provide important information. Sarcomatoid carcinomas can have well-differentiated tumour components and express epithelial markers, including CK, CK7, and CK8/18. Malignant melanomas express HMB45, MelanA, and S-100. Most leiomyosarcomas are positive for desmin, while a few cases are negative; other myogenic markers, such as caldesmon and calponin, can be combined to facilitate diagnosis. Furthermore, angiosarcomas with a cystic tubular or papillary structure and hobnail-like lining cells need to be differentiated from epithelial tumours, such as clear cell carcinoma and serous carcinoma. Small or medium-sized papillae in angiosarcomas have a fibrous axis covered by one or multiple layers of neoplastic vascular endothelial cells. Papillae in serous carcinomas are covered by multiple layers of tumour cells and can be devoid of a fibrovascular axis. Clear cell carcinomas are covered by a single layer of tumour cells, with evident interstitial collagen and eosinophilic bodies. Angiosarcomas exhibiting reticular structures or extensive bleeding also need to be differentiated from yolk sac tumour and choriocarcinoma. To distinguish whether ovarian angiosarcoma is primary or metastatic, histological and immunohistochemical findings need to be closely combined with clinical findings. Metastatic angiosarcoma usually has known primary disease foci with bilateral ovarian involvement; in addition, the surface of the ovary is usually involved and has a multilobular shape. If an angiosarcoma arises from malignant transformation of an ovarian teratoma, this is good evidence of ovarian origin.

Ovarian angiosarcoma is highly prone to relapse and has a very poor prognosis. Accurate staging is a key factor related to prognosis. When the lesion involves extraovarian tissue (stage III–IV), the prognosis is worse than when the lesion is restricted to intraovarian tissue (<stage II). In our report, the prognosis of ovarian angiosarcoma in case 1, classified as stage IA, was much better than that in case 2, classified as stage IIIB. At present, there is no uniform clinical treatment plan, and surgical resection supplemented by radiotherapy and chemotherapy is the main treatment strategy. Both anthracyclines and taxanes have significant therapeutic effects in angiosarcoma. In addition to traditional cytotoxic drugs, tyrosine kinase inhibitors have recently been used in the clinical treatment of angiosarcoma and have shown better efficacy than traditional cytotoxic drugs [29]. Tyrosine kinase inhibitors that inhibit angiogenesis reduce the tumour blood supply through inhibition of VEGF-related protein activity to achieve tumour shrinkage. Furthermore, recent studies have shown that using an anti-programmed cell death protein 1 (PD-1) antibody might be a new approach for treating angiosarcoma [30].

Angiosarcoma in the female reproductive system has a low incidence. Because of the lack of experience, it is difficult for clinicians and pathologists to make accurate diagnoses, and the challenge increases for cases with poor differentiation, rare histological types or metastatic lesions. Immunohistochemical staining for vascular markers facilitates the correct

and differential diagnosis of this tumour. Angiosarcoma has highly aggressive behaviour, and its prognosis is highly related to its clinical stage. There is limited research on genetic changes in primary ovarian angiosarcoma. Our two cases were both paraffin embedded tissues from several years ago, and the nucleic acid quality was poor, which prevented gene sequencing. More cases need to be collected for in-depth research in the future, in order to provide reference for clinical diagnosis and treatment, improve patient survival rate and prognosis.

ABBREVIATIONS

VEGF, vascular endothelial growth factor; FVIII, factor VIII; HE, Haematoxylin and eosin; CK, cytokeratin; EMA, epithelial membrane antigen; CA125, cancer antigen 125; CA 19-9, cancer antigen 19-9; AFP, alpha-fetoprotein; HCG, human chorionic gonadotropin; FIGO, International Federation of Gynecology and Obstetrics; PD-1, programmed cell death protein 1; CD, cluster of differentiation; FLI-1, friend leukaemia integration-1; G2P1, gravida 2 para 1; CT, computed tomography; AFP, alpha-fetoprotein; HCG, human chorionic gonadotropin.

AVAILABILITY OF DATA AND MATERIALS

All the generated data are included in this article.

AUTHOR CONTRIBUTIONS

DNL—manuscript writing. WYW—manuscript editing. ZYL—data collection. LL—manuscript editing.

ETHICS APPROVAL AND CONSENT TO PARTICIPATE

We applied for and received ethical approval from the Ethics Committee of West China Second University Hospital, Sichuan University (Approval No. 2022-336). Informed consent was acquired from the patient.

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CONFLICT OF INTEREST

The authors declare no conflict of interest.

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