Rare disease

Primary ovarian rhabdomyosarcoma

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Rhabdomyosarcoma (RMS) is usually considered a childhood soft tissue tumour. It normally arises within extremities, head and neck region and the genitourinary system. Common sites in the genitourinary tract include the urinary bladder, prostate and paratesticular soft tissues. It can arise at unusual sites but very rarely.¹ RMS is more common in males as compared to females and two-thirds of the cases are diagnosed in children aged 6 years or younger with a smaller incidence peak in mid-adolescence.² The annual incidence is four to seven per million children of 20 years or younger.³ Approximately 350 cases are diagnosed per year in the USA. Unfortunately, statistics of RMS are not known for Pakistan.

BACKGROUND

Embryonal rhabdomyosarcomas (RMSs) are usually localised with a favourable prognosis when compared to alveolar RMS which have the tendency to metastasise with less favourable prognosis. Pleomorphic RMS usually is an adulthood tumour and it also usually involves soft tissue of extremities. It usually has a bad prognosis. Ovarian RMSs are very rare and only 40 cases have been reported in literature till now. So a very limited data is available to predict the biological behaviour of primary ovarian RMS. Most of these ovarian RMS were either embryonal or alveolar type

with very few RMS of pleomorphic type.⁴ We are presenting a case of a young adult female who presented with lower abdominal mass and later on proved to be a case of primary ovarian RMS.

CASE PRESENTATION

A 21-year-old lady reported to gynaecologist with signs of lower abdominal mass which was slowly increasing in size and was associated with mild to moderate lower abdominal pain. There was also history of loss of appetite and weight loss. Her ultrasound and CT scan abdomen and

Figure 1 Tumour showing diffuse patternless sheets of spindle cells with intervening rhabdomyoblasts.

Figure 2 Rhabdomyoblasts showing cross striations.

pelvis showed a large mass arising from the left ovary. The tumour was localised to ovary on the initial scans. A left-sided salpingo-oophorectomy was planned and exploratory laparotomy was carried out. We received left-sided salpingo-oophorectomy, left-sided infundibulopelvic ligament and omentum. Ovary was 20×12×12 cm in size. It was intact. There was no surface growth. On serially slicing the ovary, it was found that the whole of the ovary was involved by tumour. Tumour was reaching ovarian capsule along the whole circumference. Tumour was pale to light brown in colour with multiple areas of haemorrhage and necrosis. Sections were taken from every centimetre of the tumour along with additional sections from dissimilar areas to rule out any chance of mixed tumours. Fallopian tube measured 6 cm in length and was grossly unremarkable. Infundibulopelvic ligament measured 3×2×1 cm and was showing suspicious nodular areas grossly. Omentum measuring 2×2×1 cm appeared grossly unremarkable. Sections were prepared for light microscopy.

INVESTIGATIONS

Tumour sections from ovary showed patternless diffuse sheet-like growth of proliferated spindle cells showing moderate atypia and frequent atypical mitosis (figure 1). Scattered in between spindle cells were large cells with eosinophilic abundant cytoplasm with cross striations (figure 2). Few areas of necrosis were also seen. Infundibulopelvic ligament was also involved by tumour. However, the omentum was free of tumour microscopically too. Tumour cells were positive for Desmin immunostatin (figure 3). The patient was referred to oncologist who got follow-up CT scan done. Now a small increase in size of right ovary was noted. Thinking that this may be metastatic disease, hysterectomy with right-sided salpingo-oophorectomy was planned. We received a uterus along with right-sided ovary and fallopian tube. Uterus measured 8×6×4 cm and cervix measured 3×2×2 cm. The right ovary was received in multiple fragments collectively

measuring 2×2×1.5 cm. The fallopian tube measured 3 cm in length. The cervix and uterus, on opening, were unremarkable. The right ovary revealed a heterogeneous firm area measuring 0.7×0.5 cm. The fallopian tube was unremarkable. Sections taken from the cervix, uterus and right-sided fallopian tube were unremarkable. Sections from the right ovary revealed a mature cartilage (figure 4). All of the ovarian tissue was processed but we did not find any immature area or any focus of RMS. Initially it was thought that maybe the RMS has arisen in the background of immature teratoma but, despite extensive search, no immature component was found. So we labelled this case as primary ovarian pleomorphic RMS. Cartilage found in right ovary was labelled as mature teratoma.

TREATMENT

The patient has undergone surgery twice – once initially for diagnosis and once follow-up for treatment.

OUTCOME AND FOLLOW-UP

Follow-up radiological investigations were advised by the oncologist to look for any residual disease. There was no residual disease detected on CT scan. The patient did not undergo any chemotherapy or radiotherapy as she was reluctant to do so. She is now on 6-monthly follow-up and is living a healthy life without any complications.

DISCUSSION

Primary RMS of the ovary is very rare. They usually arise in the background of teratomas, mixed mesodermal tumours or Sertoli–Leydig cell tumours. A thorough search of the tumour should be made to rule out the latter possibilities because the treatment differs. A possibility of metastasis should also be kept in mind, and a thorough search of primary RMS should also be made. So before labelling a tumour as RMS of primary ovarian origin, a thorough search of some mixed component should be made. If there is no mixed component, then a search for primary RMS

Figure 3 Tumour cells showing positivity for Desmin.

Figure 4 Right ovary showing a focus of mature cartilage.

should be made. In our case, there was no mixed germ cell, Sertoli–Leydig cell or mixed mesodermal component. In addition, a thorough clinical, radiological and skeletal survey did not reveal any primary site. Only few cases of primary ovarian RMS are reported in literature. Senger *et al*⁶ reported a case of a 2-year-old girl who presented with abdominal mass and pleural effusion. Later on, she was labelled as a case of alveolar RMS. It was also proved

by genetic studies which revealed a translocation t (2;13), which is characteristic for this tumour. Sant'Ambrogio *et al*⁶ reported a case of a 41-year-old female who was later diagnosed as a case of RMS associated with clear cell carcinoma. Allende and Yang⁷ reported a case of a 25-year-old lady who developed primary ovarian RMS with heterologous component. A review by Guérard *et al*⁸ in 1983 revealed a total of 14 cases in worldwide literature

including three paediatric patients. The review of Nielsen *et al*⁹ in 1993 revealed 13 cases which included three cases of Guérard. Six tumours involved the right ovary, three involved the left, one involved both, and laterality was not known for three patients. Microscopically, 11 tumours were embryonal RMS and 2 were alveolar RMS. Later on, Cribbs¹⁰ reported two cases of paediatric primary ovarian RMS. Chan *et al*¹¹ and Nunez *et al*¹² also reported one case each of primary ovarian RMS in their case reports.

From this limited collection of primary ovarian RMS, it is known that rhabdomyoblasts may arise in the ovary from either the uncommitted stromal fibroblasts or from fibroblasts of endometriotic stroma. The age of patients ranges between 13 months and 86 years with 60% of women older than 40 years.⁴ The survival ranged between 18 days and 15 months after diagnosis. The only hope for improved survival is combination therapy including surgery, radiation and chemotherapy.

Learning points

- From these limited reported results it is quite evident that primary ovarian RMS is a very rare tumour and RMS of any given subtype is a highly aggressive tumour. As the results are based on small number of patients and limited follow-up, they are insufficient to evaluate the effect of multidisciplinary therapy. However, it is important to correctly diagnose the case of RMS and to differentiate them from other round blue cell tumours and mixed tumours as the treatment modalities and biological behaviour are very different.

Competing interests None.

Patient consent Obtained.

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