CASE REPORT

Malignant germ cell tumour in an infant vagina

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SUMMARY

The mother of a 9-month-old female infant complained that her child was unable to pass urine at the same time noticing a mass protruding from the vaginal orifice.The infant had a single episode of vaginal bleeding.The primary concern of the mother was the inability of the daughter to micturate. Malignant germ cell tumour arising from an infant vagina is rare and accounts for about 3% of all paedriatic malignancies. These are also referred to as endodermal sinus tumours or yolk sac tumours, and are mostly the commonest form of infant vaginal malignancies encountered. A diagnosis of endodermal sinus tumour was established based on the histology and raised α-fetoprotein levels.These tumours had Schiller-Duval bodies which are primarily blood vessels surrounded by primordial germ cells and were periodic acid shift (PAS) positive diastase resistant hyaline globules which also stain positive with α-fetoprotein which is an important diagnostic feature. Tumours with high α-fetoprotein levels have a poorer prognosis. However, they respond satisfactorily to chemotherapy.

INTRODUCTION

Bleeding from the vagina in infancy and also associated with a mass is a distraughtful event to any mother. There are many underlying causes to be excuded including a foriegn body, worm infestation and malignancy. A thorough history from the mother or caretaker is essential as bleeding from a malignant tumour is a late presentation. Some of the commonest . tumours arising from an infant vagina are germ cell tumours and sarcoma botryoides. A foriegn body will cause pain and discharge besides bleeding, while worm infestation will be noticed by the mother.

Schiller described this as 'mesonephroma ovarii'since it resembles an immature glomerulus while Telum postulated two kinds of tumours with different histogenesis and age distribution,with the aggressive form occuring among the younger age groups. Scully referred this tumour as a clear cell carcinoma.

Malignant germ cell tumours are rare malignancies accounting for about 3% tumours occuring during infancy. Estimation of the α-fetoprotein is diagnostic in confirming these vaginal tumours. The prognosis worsens with tumours associated with high α-fetoprotein levels. As it is aggressive, early detection and chemotherapy are important modalities in treatment regime.

CASE REPORT

We present a case of a 9-month-old female infant whose mother complained that her child was unable to pass urine. She had also noticed a mass protruding from the vaginal orifice which was associated with a single episode of vaginal bleeding. The primary concern of the mother was her daughter's inability to micturate.

On examination, the infant was pale looking and apprenhesive. Abdominal examination revealed a soft tissue mass in the lower abdomen. A perineal examination revealed a mass palpable anteriorily which occupied the introitus and appeared haemorrhagic and protruding outside the vagina. The trans-abdominal Ultrasound and CT scans were performed.

A large sold mass filling the vagina with distension of uterine cavity and displacement of the uterus superiorly was seen on midline sagittal scan (Fig. 1). A contrast scan of the pelvis demonstrated a large heterogenous enhancing solid mass obstructing the vagina, displacing the bladder and urethra anteriorly and the rectum posteriorly (Fig. 2). The urinary bladder was compressed anteriorly as evidenced by the *in situ* balloon catheter. The mass measured 6x5cm and showed increased vascularity.

An examination under anaesthesia showed a fungating polypoid growth occupying and obstructing whole of the vagina. Biopsy taken for histopathological examination showed a possibility of sarcoma botryoides. Microscopic examination demonstrated Schiller-Duval bodies with a loose meshwork of small cystic spaces containing glomerloid-like structures lined bycuboidal epithelium and having hyperchromatic nuclei (Fig. 3 and Fig. 4). These also stained positive for α-fetoptrotein by immunohistochemistry. In our case, the mitotic index was high.There was no vascular invasion, although there were foci of tumour necrosis. No other elements of germ cell origin were identified. Serum α-feto protein was 17,158ng/ml and Human Chorionic Gonadotrophin (HCG) was less than 2U/L.

A diagnosis of malignant germ cell tumour (endodermal sinus tumour) was made.

DISCUSSION

This tumour was referred to as 'mesonephroma ovarii' since it resembled an immature glomerulus while Scully referred this tumour to as a clear cell carcinoma. The endodermal

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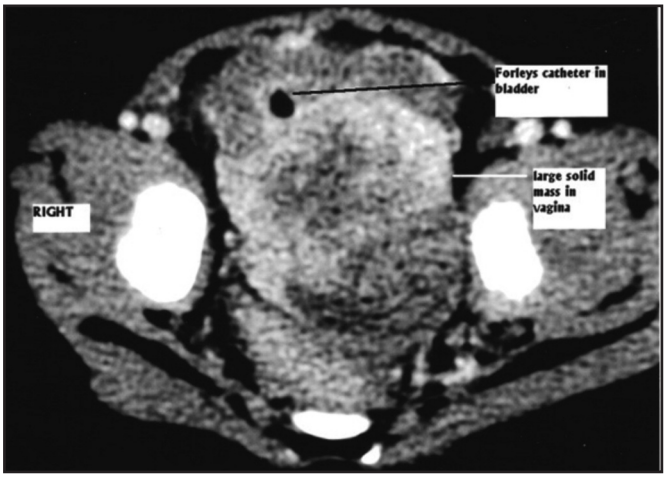

Fig. 1: Midline sagittal: Axial post IV contrast scan of the pelvis.

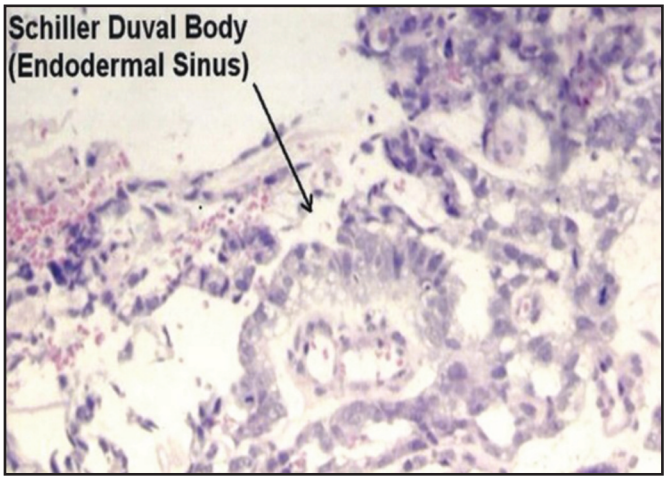

Fig. 2: Schiller-Duval body seen under H&E X20.

sinus tumour is a rare and highly malignant tumour of germ cell origin exclusively involving children. This tumour has various histopathological patterns presenting as microcysts, macrocysts, myxomatous or sarcomatoid forms. These different histological variants have no prognostic significance. The vagina is an extremely rare site for germ cell tumours which are locally aggressive and are capable of metastasis. Serum a-fetoprotein levels are useful markers for diagnosis, progression of the disease and monitoring recurrence.These tumours can be mistaken for clear cell carcinomas.

The presenting symptoms of per vaginal bleeding and the presence of a polypoidal fungating growth in the vagina of a child should guide the physician to exclude sarcoma botryoides.

The presence of Schiller-Duval bodies and PAS positive diastase resistant hyaline globules accompaied with a positive α-fetoprotein stain on immunohistochemistry confirmed the diagnosis endodermal sinus tumour. Serum α-fetoprotein is a very good immunochemical marker in patients with germ cell tumors irrespective of their location.

The serum α-fetoprotein in this case was extremely high (17,158ng/ml) indicating it of yolk sac origin and thus excluding sarcoma botryoides. A very high α-fetoprotein carries a poor prognosis. The tumour is capable of metastasis via hematogenous and lymphatic routes.

TREATMENT

Malignant germ cell tumours respond to partial vaginectomy followed by chemotherapy i.e.cisplastin, bleomycin and etoposide regime. Some have also considered to save the sexual and reproductive functions, leaving chemotherapy as the only modality of treatment. However, tumour resection alone is unsuccessful as these tumours are known to recur from any residual tumour cells. Radical surgery may be contemplated if the tumour has infiltrated the surrounding structures.

CONCLUSION

Endodermal sinus tumor is a highly malignant and rare tumor in the infant vagina. Early detection is important because of its aggressive nature and good response to chemotherapy. It is advisable that all young females with unexplained vaginal bleeding should undergo a mandatory α-fetoprotein estimation. Unfortunately we lost the patient to follow up.

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