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ORIGINAL ARTICLE

Sex Cord Stromal Tumors–Unusual Presentations

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Abstracts

Objectives The sex cord stromal tumors (SCST) account for approximately 7% of all malignant ovarian tumors. They are endocrinologically active tumors of which granulosa cell tumors are the commonest. We describe an analysis of all cases of SCST managed, in our department over the last five years and discuss two cases of unusual presentation of these tumors.

Methods Ten cases of SCST which were managed in the Department of Gynecology and Obstetrics at University College of Medical Sciences (UCMS) & associated Guru Teg Bahadur hospital, Delhi from Jan. 01 to Dec. 05 were analyzed.

Results Of the total 312 ovarian tumors which were operated in these 5 years, 73 (23.3%) were malignant, 10 cases (13.6%) out of the malignant ovarian tumors were SCST. The age of presentation varied from 4 to 70 years. Four women presented with lump abdomen as their chief complaint, 2 had postmenopausal bleeding, 2 presented with irregular bleeding per-vaginum, 1 woman had secondary amenorrhea and one young girl who was only 4½ years old developed precocious puberty. The majority of cases (70%) were granulosa cell tumors, 10% were fibromas and 20% thecomas. Maximum patients 9 (90%) were stage I at the time of surgery and only 1 case was stage IV with tumor infiltrating into omentum and gut.

Conclusion SCST tumors are rare endocrine active tumors that are essentially unilateral, benign or with a low malignant potential. Though most patients present with one or a combination of symptoms ranging from abnormal vaginal bleeding, abdominal distention and abdominal pain, the signs of Paraneoplastic syndrome (PNS) may be the first indication of the presence of an ovarian malignancy and can even facilitate its diagnosis.

Keywords Sex cord · Ovarian · Paraneoplastic · Precocious · Granulosa cell · Endometrial hyperplasia

Introduction

The sex cord stromal tumors (SCST) account for approximately 7% of all malignant ovarian tumors [1]. Majority of these tumors are of low malignant potential and are associated with favorable prognosis. SCST produce a variety of steroid hormones and the clinical presentation, ranges from precocious puberty to menometrorrhagia to postmenopausal bleeding. We describe two cases of unusual presentation of SCST which posed a diagnostic dilemma.

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Materials and Methods

Ten cases of SCST which were managed in the Department of Gynecology and Obstetrics at UCMS & associated Guru Teg Bahadur hospital, Delhi from Jan. 01 to Dec. 05 were analyzed.

Observations and Results

Of the total 312 ovarian tumors which were operated in these 5 years, 73 (23.3%) were malignant, 10 cases (13.6%) out of the malignant ovarian tumors were SCST (Table 1).

The age of presentation varied from 4 to 70 years. Four patients presented with lump abdomen as their chief complaint, 2 had postmenopausal bleeding, 2 presented with irregular bleeding per-vaginum, one woman had secondary amenorrhea and one girl who was only 4½ years old developed precocious puberty (Table 2).

The majority of cases (70%) were granulosa cell tumors, 10% were fibromas and 20% thecomas. Maximum patients 9 (90%) were stage I at the time of surgery and only 1 case was stage IV with tumor infiltrating into omentum and gut (Table 2).

Primary treatment modality was surgery in all patients followed by chemotherapy (Bleomycin, Etoposide, Cisplatin; BEP) in only 2 patients (Table 2).

Discussion

Sex cord stromal tumors are rare endocrinologically active tumors of which granulosa cell tumors are the commonest accounting for 70% of malignant SCST. Granulosa cell tumors (GCT) constitute approximately 2–5% of all ovarian malignancies. Even in our institution, GCT accounted for 70% of cases of SCST.

Though ovarian neoplasms are relatively rare in childhood and adolescence and when encountered the majority are of germ cell origin with only 5 to 7% being SCSTs. The majority of prepubertal girls present with clinical evidence of isosexual precocious pseudopuberty which was also evident in our patient who was 4½ years old with Juvenile granulosa cell tumor (JGCT). She had well developed breasts (Tanner stage III), hypertrophied labia majora and well developed axillary and pubic hair (Fig. 1). The most consistent clinical sign in patients with JGCTs is increasing abdominal girth. Young et al. [2] reported that only 2 of 113 patients in his series did not have a palpable mass. Our patient also had a large abdominal mass up to the xiphisternum. JGCTs have several behavioral characteristics which are different from Adult variety; the juvenile counterpart is characteristically aggressive and the time to relapse and death is of a limited duration. Our patient with JGCT progressed from stage Ia to stage IV within a span of 8 months and finally succumbed to her disease.

As GCTs are endocrinologically active, the patients usually present with menstrual problems. 10–15% of patients with endometrial hyperplasia may be harboring an unsuspected GCT [1]. One of our patients with Adult granulosa cell tumor presented with deranged coagulogram as the initial symptom which is a rare feature of paraneoplastic syndrome. Paraneoplastic syndrome (PNS) is characterized by clinical signs caused by certain tumors which are unrelated to the size or location of the primary tumor or its metastasis [3]. The most common PNS associated with ovarian malignancy are neurological symptoms that may be seen in as many as 16% patients. Though subacute cerebellar degeneration is the commonest paraneoplastic symptom associated with malignant ovarian tumor, defects in coagulation have also been seen.

Disseminated Intravascular Coagulation (DIC) in these patients probably occurs from tumor cell surface tissue

Table 1 Statistics

Total ovarian tumors operated	312
Malignant tumors	73
Sex cord stromal tumors	10

Table 2 Patient characteristics

	Number	Percentage (%)
Age (years)		
<20	2	20
20–40	2	20
40–60	5	50
60–80	1	10
>80		
Chief complaint		
Lump abdomen	4	40
Irregular bleeding per-vaginum	2	20
Postmenopausal bleeding	2	20
Secondary amenorrhea	1	10
Precocious puberty	1	10
Tumor type		
Granulose cell tumor	7	70
Gynandroblastoma	1	10
Thecoma	2	20
Stage of disease		
I	9	90
II	–	–
III	–	–
IV	1	10

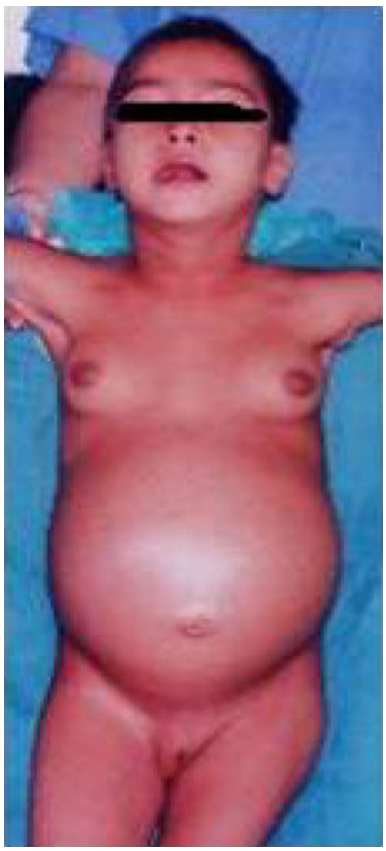

Fig. 1 4 years old child with JGCT showing precocious puberty (developed breasts, hypertrophied labia majora, presence of pubic hair) and large mass up to xiphisternum

factors, which initiate intrinsic and extrinsic pathways. Other causes could be increased serum viscosity, erythrocyte aggregation, and increased fibrogen and platelet factor 4 [4]. Rarely, intra-tumoral hemorrhage of an ovarian tumor could also lead to DIC [5]. This may have been the case in our patient.

Also, GCTs have slow indolent growth and recurrences are known even after 33 years [6]! One of our patients with GCT who was lost to follow up after primary surgery came back 15 years later with recurrences; received 6 cycles of chemotherapy followed by second look laparotomy wherein the deposits were found on gut and bladder. So, long term follow up of these patients is advocated.

In conclusion, SCST tumors are rare endocrine active tumors that are essentially unilateral, benign or with a low malignant potential. Though most patients present with one or a combination of symptoms ranging from abnormal vaginal bleeding, abdominal distention and abdominal pain, the signs of PNS may be the first indication of the presence of an ovarian malignancy and can even facilitate its diagnosis. While surgery alone is sufficient for most SCSTs, a few patients may require chemotherapy especially JGCT and Sertoli-Leydig cell tumors.

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