Ovarian Sex-cord Tumour with Peutz-Jeghers Polyp and Giant Keratoacanthoma

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Introduction

Sex cord stromal tumours represent approximately 6% of ovarian neoplasms and affect all age groups. The common types are granulosa cell tumours, fibrothecomas, and Sertoli-Leydig cell tumours. These are interesting because of their hormonal effects, which are rare for other ovarian neoplasms [1]. The sex cord tumour with annular tubules (SCTAT) is a distinctive ovarian neoplasm, the predominant component of which has intermediate morphologic features between those of the granulosa cell tumour and sertoli cell tumour. An unusual case is described consisting of large, unilateral, sex-cord ovarian tumour with annular tubules, intestinal polyps (Peutz-Jeghers), a sebaceous tumour (giant keratoacanthoma) and symptoms suggestive of hyperestrinism (persistent anovulatory cycles with dysfunctional uterine bleeding, fibroadenoma and endometriosis). Its association with Peutz-Jeghers syndrome are discussed.

Case Summary

A twenty two year old unmarried girl reported having irregular menstrual bleeding since the age of menarche for which she had been investigated and treated earlier. On diagnostic laparoscopy, she was having polycystic ovaries for which ovarian drilling was done and endometriosis was treated with depot injection provera. She improved symptomatically but developed subacute intestinal obstruction for which she was admitted in a service hospital. Clinical examination revealed an average built and nourished female, ill-looking, distended abdomen with absent bowel sounds, per rectum examination showed blood-tinged faecal matter and a palpable mass right adnexa. Radiograph of abdomen showed distended jejunal loops and air-fluid levels, contrast enhanced computed tomography (CECT) abdomen showed a solid mass right adnexa with areas of calcification and one dilated bowel loop adherent to the mass. CA-125 level was 109 U/mL. Patient was found to have lump both

breasts with firm, mobile right breast lump measuring 10 x 8 cm and left breast lump measuring 3 x 2.5cm in size. FNAC of the right breast lump was reported as an epidermal tumour.

Resection anastomosis of the gangrenous jejunal loop and resection of right ovarian mass was done. Right adnexal mass measured 6 X 5 cm, cut surface had solid and yellowish areas. Histomorphology of the ovarian mass (Fig.1) showed typical picture of sex cord tumour with annular tubules. Gross examination of the resected intestinal loop showed gangrenous gut and a polypoid mass measuring 2.5 cm in diameter. Histology of the polyp (Fig.2) confirmed a hamartomatous polyp (Peutz-Jeghers). The large lump in right breast grew rapidly, bursting spontaneously releasing a large quantity of thick cheesy material and keratotic plaque. Curettage and drainage of the remaining lump was done. Histomorphology of this lump (Fig. 3) revealed typical keratoacanthoma. Fine needle aspiration cytology (FNAC) of lump left breast was reported as fibroadenoma. Due to large size of the fibroadenoma in left breast and keratoacanthoma in right breast, the removal of these tumours would lead to loss of volume of the breasts. To make up for the proper contour of breast, silicon breast implants of 220 ml each was planned after the excision of the fibroadenoma.

Discussion

Sex cord tumour with annular tubules is a distinctive ovarian sex cord-stromal tumour. In addition to its morphologic hallmark of presence of simple and complex tubules containing eosinophilic hyaline bodies, this tumour is segregated because of its apparent high incidence among ovarian tumours associated with the Peutz-Jeghers syndrome (PJS) [2].

The authors describe a case of SCTAT associated with Peutz-Jeghers polyp as well as solitary giant keratoacanthoma in an unmarried girl. This patient had typical Peutz-Jeghers polyp and SCTAT but diagnosis of PJS could not be made since the hamartomatous

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Fig 1 : Sex-cord stromal tumour with annular tubules having fibrothecomatous cellular stroma containing solid tubules with hyaline homogenous material in the center (x 40).

Fig 2 : Peutz- Jegher's polyp having central core with branching smooth muscle strands lined by normal looking mucin-secreting epithelial cells (x 10).

polyp was not associated with mucocutaneous pigmentation or family history. Giardiello, et al [3], defined a definite diagnosis of PJS as histopathologic confirmation of hamartomatous polyps and the presence of at least two of the following criteria; family history, hyperpigmentation or small bowel polyposis. Patients with PJS have an increased risk for breast, gastrointestinal and female genital tract cancers. Most female PJS patient will have ovarian involvement by SCTAT. In women with PJS, SCTATs are usually multifocal, bilateral, small, calcified and benign, whereas the sporadic forms of SCTAT are unilateral, often large (palpable) and malignant in 20% of cases. Approximately 36% of SCTATs are associated with PJS [4].

In this patient no lymphatic spread of SCTAT was detected. SCTAT is considered as a tumour with low-grade malignancy and retroperitoneal lymphatic metastasis is thought to be an important pathway of spread. In cases of SCTAT occurring in PJS patients, all the reported cases with one exception have behaved in a benign fashion [5]. Among the cohort of patients

Fig. 3 : Histological findings of sporadic keratoacanthoma with marked epithelial proliferation and a crater filled with a large keratin plug (x10).

with PJS, clinically malignant behaviour in SCTAT occurs with a much higher frequency and metastases developed in seven (15%) of 47 patients in the Young et al series [4]. In either case, SCTAT may be associated with significant morbidity, such as precocious puberty, bleeding anomalies or premature ovarian failure. In further support of the dichotomy between the PJS-associated SCTAT and the non-PJS-associated cases, are the molecular findings recently reported by Connolly et al [6]. Mutations in the PJS-associated gene *STK11*, which maps to 19p13.3 were found in two PJS-associated SCTAT cases and in none of five non-PJS-associated SCTAT cases [6].

Our patient also developed giant keratoacanthoma of the breast which rapidly increased in size and burst spontaneously. Giant keratoacanthoma is a rare clinical variant of solitary keratoacanthomas [7] and is known to grow rapidly. Since keratoacanthoma or keratoacanthoma-like sebaceous adenomas qualify as a component, the diagnosis of Muir-Torre Syndrome (MTS) was considered in this case of SCTAT. Keratoacanthomas commonly occur in the MTS [7]. MTS is a rare autosomal dominant condition in which multiple primary malignancies occur with a sebaceous gland tumour. Criteria for the diagnosis of MTS includes the presence of at least one sebaceous adenoma, sebaceous epithelioma, or sebaceous carcinoma and one internal malignancy. Keratoacanthoma, squamous cell carcinoma and multiple follicular cysts are included in the cutaneous tumours.

Colorectal cancer is the commonest initial visceral neoplasm in MTS, while genito-urinary cancers occur in 24% of cases. Carcinomas of the endometrium, ovary, breast, parotid, upper gastrointestinal tract, larynx and haematological malignancies are also associated with MTS. As in the Lynch II syndrome, cancers occur at a

relatively young age. Colonic polyps are found in more than 25% of MTS patients and are common in patients with colorectal carcinoma. Early recognition of MTS in patients with sebaceous gland tumours should facilitate early detection of subsequent malignancies [8]. The sebaceous gland tumours (a marker for MTS) are rare in the general population and finding one should prompt a search for occult malignancy. Cohen et al [9], suggested that a search for internal malignancy should be done in patient including the family members when ever an MTS associated sebaceous gland tumour is diagnosed.

Conflicts of Interest

None identified

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Answer to Pictorial Quiz

Chromoblastomycosis

The histopathological study of sections stained with haematoxylin and eosin showed hyperkeratotic epidermis with acanthosis and a diffuse infiltration of lymphocytes, histiocytes, eosinophils and plasma cells in the dermis along with neutrophilic microabscesses containing brown sclerotic bodies of pigmented fungal infection. Culture of the tissue grew *Cladosporium carrionii*.

Discussion

Chromoblastomycosis is a chronic fungal infection of the skin and subcutaneous tissue caused by traumatic inoculation of a specific group of dematiaceous fungi usually *Fonsecaea pedrosoi*. It occurs commonly in tropical and subtropical countries, caribbean regions, Africa, Australia and Japan. The lower extremities are commonly affected often following superficial trauma and a locally spreading verrucous plaque or a solid nodule develops at the site of inoculation which is generally refractory to treatment. Rarely involvement of the penile shaft, vulva, ala of nose, pleural cavity, ileocaecal region, laryngotracheal area and tonsils has been reported [1].

Four different genera are known to cause chromoblastomycosis namely *Fonsecaea pedrosoi*, *Phialophora verrucosa*, *Cladosporium carrionii* and *Fonsecaea compacta* however rare cases due to *Rhinocladiella aquaspersa* and *Exophiala species* have also been reported. Approximately 70% of cases are seen in males as they are more commonly involved in agricultural work and

prone to inflict injuries on themselves.

The verrucous forms need to be differentiated from tuberculosis verrucosa cutis and from squamous cell carcinoma which may develop in long standing lesions of chromoblastomycosis. There are also reports of chromoblastomycosis masquerading as palmoplantar psoriasis and sporotrichosis [2,3].

The diagnosis is based on KOH examination, identification of the organism in haematoxylin and eosin sections, culture of the organism that reveals slow growing green to black colonies and microscopic appearance of conidia. Early stages of chromoblastomycosis are treated with surgical excision, electrodesiccation or cryosurgery. Advanced cases may require systemic antifungals for long periods of time. Itraconazole and terbinafine are the drugs of choice [4].

Conflicts of Interest

None identified

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