An Unusual Presentation of a Sertoli Cell Ovarian Tumor in a Prepubertal Patient

Silvia Bernardo, Erica Makin, Olivia McKinney and Saira Haque*

Department of Radiology, King’s College Hospital, UK

Abstract

Sertoli-Leydig tumors belong to the category of ovarian tumors derived from specialized gonadal stroma. Clinical features include endocrine symptoms, fatigue, chronic abdominal pain, abdominal distension and symptoms related to localize mass effect. We describe a case of pure Sertoli cell tumor in a 6-year-old girl who had an atypical presentation initially with intermittent right hip pain radiating to the groin. We will also highlight key Magnetic Resonance (MR) imaging findings which have not been well documented in the literature in prepubertal children.

Introduction

Ovarian tumors are seldom seen in children and Sertoli-Leydig cell tumors account for less than 0.5% of these cases [1]. These are the most common virilizing ovarian tumor, which usually occur in young women. Approximately 30% of cases are associated with androgenic activity. Precocious puberty is rarely reported in prepubertal girls. If the Sertoli cells predominate, the effects of excess estrogen may be seen; in a tumor composed primarily of Leydig cells, the androgenic effects predominate. A case of a pure Sertoli cell tumor in a 10-year-old female without symptoms of isosexual pseudoprecocity has been reported [2]. We report a second case in which the patient initially presented with intermittent unilateral right sided hip pain and then developed intermittent lower abdominal pain over a six-month period. The diagnosis was made after presentation to the emergency department with an acute episode of severe abdominal pain and vomiting.

Case Presentation

A 6-year-old Afro-Caribbean female patient presented to her primary care physician with intermittent, dull, right sided hip pain. The patient was born at 31 weeks and required treatment for respiratory distress initially with surfactant and was self-ventilating within 48 h. At one year of age, the child was noted to have pubic hair, although hormonal studies at the time were reported to be normal. No other significant past medical history. The ultrasound examination of the hip and groin region was normal. The child represented two months later to the emergency department with lower abdominal pain. A diagnosis of a urinary tract infection was made after a urine sample taken on admission showed an increase in the leukocyte count and a trace of protein. The patient was given a course of oral antibiotics for seven days resulting in partial resolution of symptoms. Three months later, the patient presented to the emergency department with severe abdominal pain and bilious vomiting. On clinical examination, there was a palpable abdominopelvic mass extending up to the level of the umbilicus. There was a small amount of pubic hair noted. No axillary hair, breast development, uterine bleeding, clitoral enlargement, vaginal discharge or advanced skeletal growth. An ultrasound demonstrated a large mass arising from the right ovary containing solid and cystic components with internal thick septations. The left ovary and uterus were normal. A small amount of pelvic free fluid was seen. Except for a mildly elevated lactate dehydrogenase of 377 IU/L (normal value <240 IU/L), all tumor markers were normal. This included beta human chorionic gonadotropin <2 IU/L, alpha-fetoprotein <2 kIU/L, cancer antigen 125 was 11 kU/L, carcinoembryonic antigen <2 ng/L, carbohydrate antigen 19-9 was 19 kU/L and inhibin B was 159 ng/L. Her serum hormonal profile including the estrogen, estradiol and progesterone levels were normal.

MR demonstrated a large, complex solid mass (9 cm × 7 cm × 6 cm) with engorgement of the vessels (Figure 1a and 1b). The mass was of low to intermediate signal on T2-weighted imaging and marked contrast enhancement (Figure 1c and 1d). A subsequent laparotomy was performed which revealed a mobile mass enveloping the right ovary, with associated edema of the right fallopian tube. A right oophorectomy with sparing of the fallopian tube was performed. Peritoneal fluid and omental biopsy was taken for cytology and
histology respectively, which confirmed no evidence of malignancy. Macroscopic histology showed that the tumor was solid and nodular, weighing 200 gr. On sectioning, the parenchyma demonstrated several microcysts containing gelatinous and mucoid material. The solid areas showed tumor composed of tubules and trabeculae interspersed by thin fibrous bands (Figure 2). The cytoplasm ranged from pale to eosinophilic. No normal ovarian tissue was identified surrounding the tumor. There was no evidence of lymphovascular invasion. The definitive histology confirmed the diagnosis of a completely excised large purely Sertoli tumor of the right ovary.

**Discussion**

Ovarian sex cord-stromal tumors develop from the dividing cell population that would normally produce cells that support and surround the oocytes, including those that produce ovarian hormones. Ovarian sex cord-stromal tumors account for 10% to 20% of all pediatric ovarian tumors and 5% to 10% of ovarian neoplasms in all age groups. They frequently occur in the pediatric age group, particularly in girls younger than 15 years [3]. These tumors are classified into four types: Granulosa-stromal cell tumors, Sertoli-stromal cell tumors, sex cord-stromal tumors of mixed or unclassified cell types, and steroid cell tumors. In children and adolescents, granulosa cell tumor and Sertoli-Leydig cell tumor commonly occur. Pure Sertoli cell tumors do not contain the immature neoplastic stroma found in the Sertoli-Leydig tumors. Pure Sertoli cell tumors are rare, accounting for 4% of Sertoli-Leydig cell tumors [6]. There are over 350 reported cases of Sertoli cell tumors, in children and adults, and there is limited knowledge of its prognosis. Among them, only seven cases of Sertoli cell tumors in prepubertal patients have been reported in the literature. Six of these females presented with isosexual pseudoprecocity [6]. Sertoli-Leydig cell tumors are classified into well-differentiated, moderately differentiated, poorly differentiated and retiform subtypes [7]. All except the well-differentiated subtype may contain heterologous elements. Most of these tumors are diagnosed as early stage and low-grade malignant tumors and the prognosis is usually good [8].

Sertoli-Leydig cell tumors tend to be unilateral and can be solid, solid and cystic, cystic or papillary. It typically appears as a predominantly solid mass with peripheral or intratumoral cysts or as a cystic mass with solid mural portions [9]. A well-demarcated enhancing solid mass lesion which contains multi-cystic areas of variable size is seen on MR imaging. These cystic areas are secondary to heterologous elements. On T2-weighted MR imaging, the solid components show variable signal intensity, depending on the extent of fibrous stroma; low signal intensity on T2-weighted imaging is indicative of greater fibrous stroma content. The differential diagnosis of ovarian tumors with enhancing solid portions also includes ovarian cystadenofibromas, struma ovarii and sclerosing stromal tumors (benign subtype of ovarian sex cord stromal tumors) [10].

Adnexal torsion was considered as a cause of the intermittent
nature of the lower abdominal pain, following the diagnosis of the large ovarian mass on the ultrasound. Torsion of the ovarian pedicle produces circulatory stasis, which initially obstructs venous low-pressure flow and subsequently higher-pressure arteries become constricted which leads to massive ovarian edema. Imaging features include an enlarged ovary, an ovarian mass with cortical follicles, a twisted vascular pedicle, and ipsilateral fallopian tube enlargement, wall thickening of the adnexal mass, ascites and uterine deviation to the twisted side [11]. In this case, MR imaging demonstrated multiple enlarged vessels adjacent to the right ovary and pelvic sidewall although there were no other ancillary features to suggest adnexal torsion. Though it should be noted that torsion may occur concurrently with adnexal tumors.

The staging of ovarian tumors on imaging is important as it plays a key role in patient management, particularly the surgical approach to excision and ultimately planning a treatment regimen. Ovarian tumors are staged based on the extent of spread of the tumor (Table 1). A stage I tumor is contained within the ovary. Stage II has spread to nearby organs and stage III indicates unilateral or bilateral ovarian involvement and the following (a) spread beyond the pelvis to the peritoneal cavity (b) lymphatic spread. Stage IV involves distant metastatic spread, particularly liver and lung metastases. This case showed features of stage I disease, with no side wall spread in theatre. The survival rate for patients with Sertoli cell tumors is higher than 90%. The treatment of choice for stage IA tumors is surgical excision with no chemotherapy [12].

In summary, we outlined the atypical clinical presentation of a purely Sertoli tumor in a prepubertal patient, initially presenting with unilateral hip pain radiating to the groin. We have also outlined key MR imaging findings including predominantly low signal on T1-weighted imaging, intratumoral cystic change on T2-weighted imaging and marked contrast enhancement.

References