Primary Strumal Carcinoid of the Ovary- A Rare Entity

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Abstract: Carcinoid Tumors of the ovary are very rare tumors. They can be either primary or metastatic in origin. Primary ovarian carcinoid tumors can be either a part of a mature cystic teratoma or a pure tumor. We report a case of primary strumal carcinoid tumor in a female patient aged 29 years who presented with chronic constipation and abdominal distension.

Keywords: strumal carcinoid, ovarian carcinoid, mature cystic teratoma

1. Introduction

Primary Carcinoid Tumor of the ovary is a relatively uncommon tumor commonly occurring in young adults [1]. Histologically they can be subdivided into 5 categories: Insular, Trabecular, Strumal, Mucinous and Mixed types[1]. Of all these varieties insular are the most common type and strumal variety is the least common. Many of the primary carcinoid tumors are associated with symptoms of Carcinoid syndrome, irrespective of the size of the tumor. In the past one decade we have reported a single case of Primary Ovarian Strumal Carcinoid in a female patient aged 29 years and we are presenting it.

2. Case Report

A 29 year old female patient, multiparous, married since 7 years attended the surgical Outpatient department with non specific symptoms such as chronic constipation and abdominal distension since 1 year. The symptoms were waxing and waning. Routine examination and investigations were performed. Ultrasonography of the abdomen revealed a 10X7X5 cm solid mass in the right ilial fossa suggestive of an ovarian malignancy. Hence the patient was referred to the gynecological department who performed a unilateral salpingo-oophorectomy and removed the mass. The mass was sent to the Department of pathology for histopathological examination.

The specimen was routinely processed with 10 % formalin for fixation. Gross examination revealed a solid mass of maximum dimension of 9 cm, with a smooth external surface. Cut section revealed a solid, light tan to yellowish glistening surface with a small cyst of 2 cm diameter. Areas of hemorrhages were also noted (Fig 1).

Bits were taken from representative areas and routinely processed and sections stained with Hematoxylin and Eosin was examined microscopically.

Microscopic examination revealed a tumor composed on intimate mixture of thyroid follicles containing colloid and trabeculae of neoplastic neuroendocrine cells, separated by thick fibrous septae, resembling trabecular carcinoid.individual cells showed elongated nuclei with finely dispersed chromatin in these areas. There were small areas revealing hemorrhages and necrosis. There was increased mitotic activity noted in the carcinoid areas. Hence, with these features a histopathological diagnosis of a Strumal Carcinoid was made (Fig 2).

Immunohistochemistry was also performed. The strumal component was positive for CK7 and the carcinoid component was positive for chromogranin confirming the diagnosis.

The patient has been followed up till date with no recurrence and is clinically well.
3. Discussion

We report this case for its rarity and its presenting symptoms. Primary strumal carcinoid of the ovary is a very rare entity. Around 60 cases have been reported in the world literature with probably quite a few unreported cases [2]. The age distribution varies from 21 to 77 years. The tumor is usually associated with non specific complaints. However, there are a few cases which presented with features of Carcinoid syndrome [3].

Most often these tumors occur as a component of a mature cystic teratoma, where they occur as a yellowish nodule. However, though less common, pure solid forms are also seen.

Strumal carcinoids are ovarian neoplasms which have combined features of both struma ovarii and ovarian carcinoid, with either of them being more predominant. Carcinoid component is most commonly the trabecular variant. However, rare cases with mucinous carcinoid component have been reported [4].

The origin of both the components of the tumor is still debatable. Some authors have the opinion that strumal carcinoid is a carcinoid tumor with areas representing thyroid tissue. This has been disproved by others by demonstrating expression of thyroglobulin[5]. Some authors suggest that the carcinoid component has its origin from thyroid C cells while others consider it as a separate entity [5].

Occasional cases have also demonstrated a papillary microcarcinoma in strumal component of the tumor [6]. Other teratomatous elements can be a component of the tumor and are reported in many cases.

Immunohistochemical studies are of value in cases of doubt of the diagnosis. The strumal component of the tumor shows expression of thyroglobulin, TTF-1 and CK7 and the carcinoid component shows expression of chromogranin and synaptophysin which is in concordance to their histological features [7]. Strumal carcinoids usually have a good prognosis with a benign course, unless associated with metastasis, which by itself is a very rare phenomenon [8].

References