Primary testicular carcinoid tumors are extremely rare and account for less than 1% of all testicular tumors. We recently experienced a case of primary carcinoid tumor of the testis in a 50-year-old man who presented with scrotal swelling. The ultrasonographic findings were evaluated and compared with those of previous reports.

Index words: Carcinoid
Testicular tumor
Ultrasound (US)

Carcinoid tumors of the testes are very rare. Since Simon et al. reported the first case of primary testicular carcinoid tumor, about 70 carcinoid tumor cases have been recorded in the literature. However, no reports on the imaging findings of primary testicular carcinoid tumor have been published in the Korean literature.

We report a case of primary testicular carcinoid tumor along with a description of the ultrasonographic findings.

Case Report

A 50-year-old man presented with left scrotal swelling which he had been suffering from for three months. The patient denied having any other symptoms. He was referred to our hospital by a local clinic for further evaluation. Physical examination revealed a palpable non-tender hard mass in the left testis. Scrotal ultrasonography showed an isoechoic solid mass with a peripheral hypoechoic rim and small anechoic cystic areas in the left testis, but no calcification was seen (Fig. 1A). Increased flow signals surrounding the intratesticular mass were seen on the Doppler image (Fig. 1B). The right testis was normal in both the physical examination and ultrasonography. We suspected a non-seminomatous germ cell tumor of the left testis and, although secondary intratesticular cancer is uncommon, the presence of a secondary malignance could not be excluded.

The levels of serum testicular tumor markers such as $\alpha$-fetoprotein, $\beta$-human chorionic gonadotrophin and lactate dehydrogenase were normal. However, the level of urine 5-hydroxy-indole acetic acid was not checked. Computed tomography scans of the chest, abdomen and pelvis were unremarkable. The patient underwent left orchiectomy.

The gross specimen consisted of an ovoid, solid, whitish-yellow mass, measuring $3.2 \times 2.4 \times 2.0$ cm replaced by tumor except for the peripheral viable area (Fig. 1C). The nodular surface showed small cysts in the upper portion and spotted hemorrhage. Immunohistochemically, the tumor cells were positive for cytokeratin, NSE and chromogranin (Fig. 1D). However, PLAP immunostaining was negative. The electron microscopic study showed numerous dense-core neurosecretory granules in the cytoplasm of the tumor cells (Fig. 1E). The level of 5-hydroxy-indoleacetic acid in the urine collected fol-
following the operation was within the normal range.

Discussion

Primary testicular carcinoid tumors are extremely rare, and most patients with this disease present in their fifth decade of life [1]. The origin of carcinoid tumors is argentaffin or Kulchitsky cells, which are usually located within the crypts of Lieberkuhn [2].

Testicular carcinoid tumors have been divided into three subgroups, viz. primary testicular carcinoid tumors, carcinoid tumors associated with teratoma and carcinoid tumors metastatic to the testis. Even though the latter two groups are relatively unusual, the most important differential diagnosis is metastatic carcinoid tumor, and the differentiation between primary and metastatic carcinoid tumor is difficult, both clinically and morphologically [3]. To establish the diagnosis of a primary carcinoid tumor, the existence of primary lesions elsewhere in the body must be excluded [4]. The majority of carcinoid tumors originate from the gastrointestinal tract, especially the appendix and ileocecal region. CT of the abdomen and pelvis should be performed to exclude the existence of primary tumors in this region and lymphatic spread. The second most common origin of carcinoid tumors is the lung; therefore, a plain radiograph of the chest or chest CT is helpful to rule out the presence of bronchial carcinoid tumor [1, 5, 6].

A painless mass and prominent testicular enlargement are common clinical findings. Other symptoms including carcinoid syndrome are rare in patients with prima-
ry testicular carcinoid tumors.

Ultrasonography is the primary imaging modality in the evaluation of testicular tumors. Only a few cases (of primary testicular carcinoid tumor?) were reported previously (2, 3, 7). In these cases, the US features included a solid well-circumscribed hypo or iso-echoic intratesticular mass with dense calcification. Grunshaw et al. reported dense focal nodular calcification at the periphery of the mass. Calcifications located within the testicular parenchyma are found in germ cell tumors, especially in teratoma, embryonal cell carcinoma and granulomatous orchitis (8, 9). Microcalcifications due to teratoma represent calcified cartilage and/or bone fragments (9). However, the calcification in primary testicular carcinoid tumors may be dystrophic (2).

Some cases of primary testicular carcinoid tumor showed small cystic or necrotic foci within the mass (7, 8). In our case, even though there was no focal calcification, the ultrasonic appearances of the testicular carcinoid tumor showed small cystic areas within a hypoechoic intratesticular mass.

Because of the generally favorable prognosis, the management of patients with a pure primary testicular carcinoid tumor usually includes inguinal orchietomy for locally confined tumors and the surgical resection of isolated metastatic foci (2). The overall incidence of metastasis is about 11% (1). A review of the literature showed that the tumor size and the presence of carcinoid syndrome are features associated with a malignant course.

The elevation of urinary 5-hydroxyindoleacetic acid (5-HIAA) and serum serotonin levels following orchietomy often suggests the possibility of a metastatic tumor (1, 10).

Although their ultrasonographic findings are not specific, testicular carcinoid tumors should be included in the differential diagnosis of a hypo- or isoechoic solid intratesticular mass containing calcification and small cystic or necrotic foci and, once diagnosed, it is important to carefully assess whether the tumor is primary or metastatic.

References