

Case Report



Paratesticular Fibrous Hamartoma In An Adult: Case Report and Review of the Literature

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ABSTRACT

Fibrous hamartoma (FH) is a focal malformation resembling a neoplasm, composed of an overgrowth of mature cells and tissues that normally occur in the affected area. But they are characterized by disorganization. FHs are usually seen at childhood and located in head and neck region, gastrointestinal system and lung. It is not reported previously in the paratesticular region and in adults. We reported a 47 years-old man with right testicular mass that was diagnosed as paratesticular fibrous hamartoma. In English literature, this is the first case of a fibrous hamartoma originating from paratesticular region in adults. We aim to discuss this case in light of the literature. ©2008, Fırat University, Medical Faculty.

Key words: Fibrous hamartoma, paratesticular tumor, intrascrotal mass

ÖZET

Erişkinde Paratestiküler Fibröz Hamartom: Olgu Sunumu ve Literatür Taraması

Fibröz hamartom (FH), normal olarak bir dokuda bulunan matür hücre ve doku elemanlarının aşırı büyümesiyle oluşan, neoplazmi taklit eden fokal bir malformasyondur. Ancak bu dokuların düzensiz çoğalmasıyla karakterizedir. FHlar, genellikle çocukluk çağında görülür ve baş-boyun bölgesi, gastrointestinal sistem ve akciğerlerde ortaya çıkar. Ancak daha önce erişkinde paratestiküler yerleşim bildirilmemiştir. Sağ testiküler kitlesi olan 47 yaşındaki erkek hastaya paratestiküler fibröz hamartom tanısı koyduk. Bu, İngilizce literatürde erişkinde paratestiküler yerleşimli ilk fibröz hamartom olgusudur. Olguyu literatür eşliğinde tartışmayı amaçladık. ©2008, Fırat Üniversitesi, Tıp Fakültesi

Anahtar kelimeler: Fibröz hamartom, paratestiküler tümör, intraskrotal kitle

Hamartoma is a focal malformation resembling a neoplasm, composed of an overgrowth of mature cells and tissues that normally occur in the affected area. However, these mature cells and tissues are disorganized and one of the cell or tissue type is predominant (1). To our knowledge, paratesticular fibrous hamartomas has not been reported in adults. Hence this is the first case reported in the English literature.

CASE REPORT

A 47-years-old patient suffered with right testicular mass was admitted to department of urology, Fırat Medical Center. In the clinical history, the patient has the testicular mass since 20 years. The patient had been operated for right inguinal hernia 9 days ago, before admitting to our clinic. The major symptoms of the patient were testicular enlargement and pain.

In the color doppler ultrasonography examination, right testis was in normal sizes and its echo was minimally decreased compared to the left testis. The echo of epididym was decreased and its blood flow was increased. Testicular perfusion in the right was minimally increased. Thickness of the scrotal skin was clearly increased at the right side. On the right side, a mass extending from inguinal channel to the scrotum about 8x7 cm was seen.

There was cystic areas, echogen particles similar hemorrhagic focus and hypoechogen foci resembling inflamed connective tissue. There was a mass extending from right inguinal canal to right hemiscrotum. Radiological diagnosis was as an abscess or hemorrhage (epididymoorchitis?). Testicular tumor's markers and results of hematological and biochemical analysis were normal. Right orchiectomy was performed.

In macroscopic examination, the mass which was 7 cm in maximum diameter and had focal hemorrhage areas was composed of fibrous and adipose tissue and pushed normal testicular tissue (Figure 1).

In microscopic examination, the tumor showed disorganized mature tissue that composed of thick-walled vessels, fibrous, muscular and adipose tissue (Figure 2-3).

Additionally, chronic inflammatory infiltration, foreign body reaction and cholesterol clefts were seen in the disorganized tissue (Figure 4). By these findings, it was diagnosed as paratesticular hamartoma

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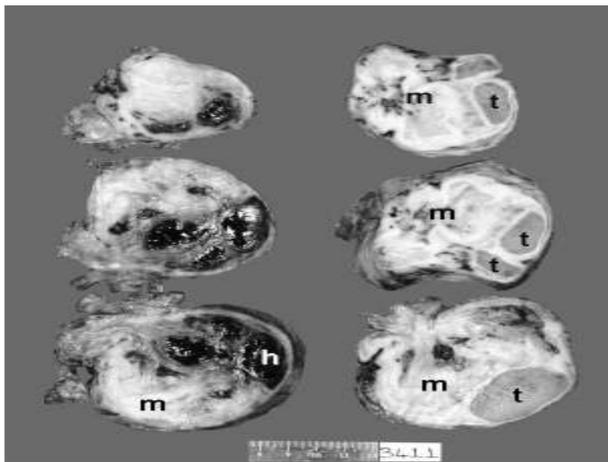


Figure 1. Slices of the orshiectomy specimen show paratesticular mass (m) that pushing testis (t) and contain hematoma (h) focuses.

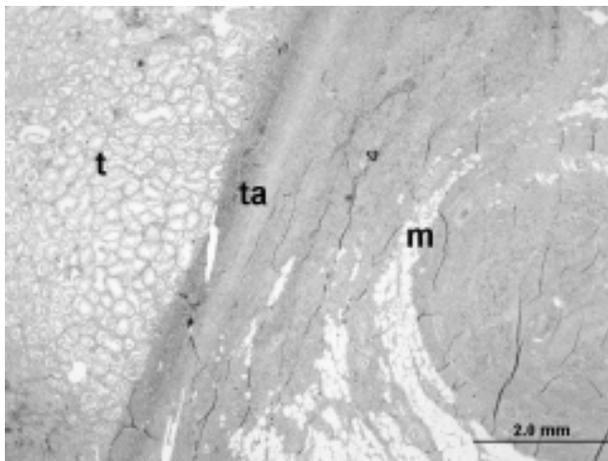


Figure 2. Seen the paratestiküler mass (m) that contain disorganized soft tissues (t: testis, ta: tunica albuginea) (H&E, x40)

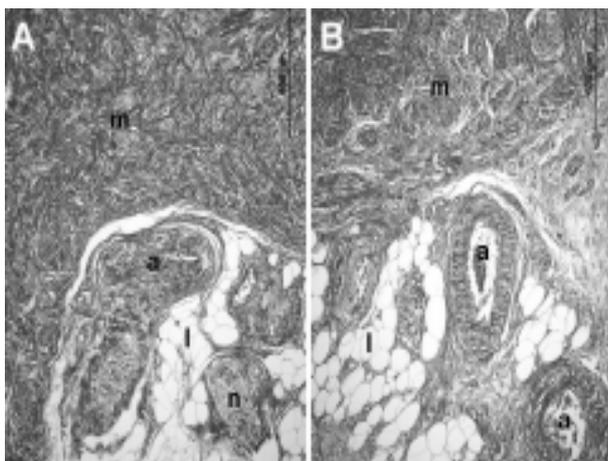


Figure 3. A and B; Paratesticular mass composed of irregular muscle bundles (m), arteries (a) with thick wall, nerves (n) and adipous tissue (l) (H&E, x100)

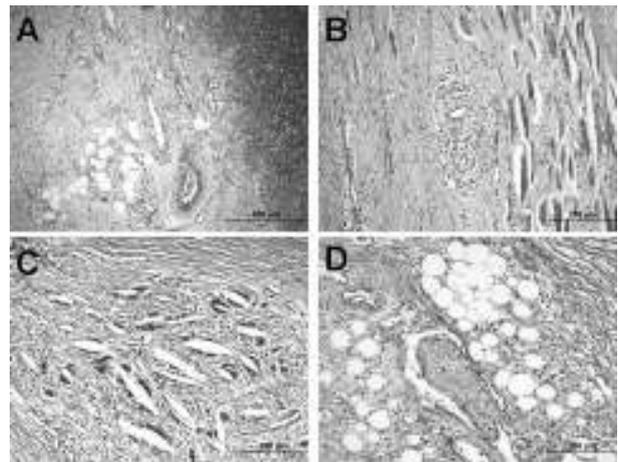


Figure 4. Postoperative changes: **A;** hematoma area (H&E, x100), **B;** inflammation and fibrosis that infiltration between muscle bundles (H&E, x200), **C;** foreign body reaction and cholesterol clefts (H&E, x200), **D;** fat necrosis and histiocyte infiltration (H&E, x200)

DISCUSSION

The hamartoma word is originated from “hamartia” in Greek alphabet and meaning “scribal error or mistake”. Hamartomas may be seen anywhere in the body and are often seen in infancy and childhood. Therefore, it has been believed that these lesions are developmental aberrations (2). It is often seen in the head and neck region (especially around ear), gastrointestinal system and lungs and rarely seen in corpus cavernosum, larynx, urinary bladder, hypothalamus and retina (3-7). To date, paratesticular fibrous hamartoma in adults has not been reported in the English literature.

Tumors of the spermatic cord and paratesticular region are uncommon. The paratesticular area is a complex anatomical area which includes the contents of the spermatic cord, testicular tunics, epididymis and vestigial remnants, such as the appendices epididymidis and testis. Histogenetically, this area is composed of a variety of epithelial, mesothelial and mesenchymal elements. Neoplasms arising from this region therefore form a heterogeneous group of tumors with different behavioural patterns and have a wide differential diagnosis. The clinical presentation is almost always a mass or swelling, which may or may not be painful and is occasionally accompanied by hydrocele. These clinical findings do not help to distinguish a benign tumor from a malignant tumor. Paratesticular tumors can present at any age, which may sometimes give a clue to the histological diagnosis (8).

Lioe et al. (8), in 36 years period, reported 85 paratesticular tumors in which 66 (78%) were benign lesions and the remaining 19 were malignant. The most common benign lesion was the adenomatoid tumour. Lipomas are also seen frequently (9,10). Other much rarer benign lesions included leiomyoma, haemangioma, fibroma, neurofibroma and papillary mesothelioma. While lipomas are seen at a large interval of age (2-71 age), adenomatoid tumors are often seen between 20 to 50 ages and rarely seen in childhood. Sizes of these tumors are variable. While adenomatoid tumors are rarely over 10 cm, diameters of lipomas and malignant tumors are usually over 10 cm (8).

In another study (11), twenty two patients with tumours or tumour-like conditions of the paratesticular region were evaluated over a 5 years period. Of these, 16 (73%) were benign with only one true neoplasm (papillary cystadenoma). The tumor-like conditions were comprised of 6 cases of adenomatous hyperplasia of epididymis, 4 cases of spermatic granuloma, 2 cases each of spermatocele and nodular-fibrous proliferation and one mesothelial cyst (11).

To date, paratesticular hamartoma cases have been reported in English literature. The first case was reported by Srigley and Hartwick, in a two years old child (1). They described that the tumor consisted of a disorganized cluster of tubules embedded in a loose connective tissue stroma and tubules were lined by cells that were cytologically similar to normal rete testis. This tumor is called as hamartoma of rete testis and is morphologically different from fibrous hamartoma.

Histopathologic features of our case were compatible with fibrous hamartomas described in the literature. On

contrary to the literature, in our case, focally dense chronic inflammatory reaction, lymphoid aggregations, foreign body reaction and cholesterol clefts were seen. We believe that the most of these changes are developed secondary to lipid extraction from cells, due to chronic trauma or surgical operation.

The second case is reported in childhood by Jimenes and et al. (12) and was diagnosed as "paratesticular fibrous hamartoma". However, we could not get information about morphological features of this case. According to these reports, our case is the first paratesticular fibrous hamartoma in adulthood.

Finally in the differential diagnosis of the paratesticular tumors, paratesticular fibrous hamartomas should have been taken into consideration. These tumors may be confused with malignant tumors of testis and paratesticular region. Preoperative diagnosis of the paratesticular hamartomas prevents unnecessary orchiectomy.

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