CASE REPORT

INTRASCROTAL EXTRATESTICULAR NEUROFIBROMA: A CASE REPORT

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ABSTRACT

Neurofibroma is a benign tumor of the nerve sheath, which is considered to originate from the Schwann cell. Since neural tissues are found throughout the body these tumors can occur in a variety of sites. Intrascrotal extratesticular localization of neurofibromas have been reported extremetely rarely in the literature. A 45-year-old man admitted to our clinic with a four year history of gradually enlarging mass and scrotal discomfort in the left hemiscrotum. With a preoperative diagnosis of testicular or paratesticular tumor, exploration and frozen examination were performed. The pathological diagnosis of the lesion was neurofibroma of the paratesticular region. Although a relatively rare disease, intrascrotal extratesticular neurofibroma should be considered in the differential diagnosis of testicular and paratesticular malign tumors.

Keywords: Neurofibroma, Intrascrotal, Extratesticular

İNTRASKROTAL EKSTRATESTİKÜLER NÖROFİBROMA: OLGU SUNUMU

ÖZET

Nörofibroma Schwann hücrelerinden kaynaklandığı düşünülen sinir yapılarının benign tümörüdür. Nöral dokular vücudun hemen heryerinde bulunduğu için bu tümörler çeşitli lokalizasyonlarda görülebilmektedirler. Literatürde oldukça nadir olarak intraskrotal ekstratestiküler lokalizasyonlu nörofibroma bildirilmiştir. Kırkbeş yaşında erkek hasta kliniğimize sol skrotumda rahatsızlık ve sol testiste son dört yıldır yavaş yavaş büyüyen şişlik yakınması ile başvurdu. Preoperatif olarak testiküler ya da paratestiküler tümör ön tanısı ile eksplorasyon ve frozen inceleme yapıldı. Kitlenin patolojik incelemesi patolojinin intraskrotal ekstratestiküler nörofibroma olduğunu ortaya koydu. Oldukça nadir görülmelerine rağmen ekstratestiküler intraskrotal nörofibromalar malign testiküler ya da paratestiküler tümörlerin ayırıcı tanılarında gözönünde bulundurulmalıdırlar.

Anahtar Kelimeler: Nörofibroma, İntraskrotal, Ekstratestiküler

INTRODUCTION

Neurofibroma is a benign tumor of the nerve sheath, which results from an abnormal overgrowth of Schwann cells[1]. Since neural tissues are found throughout the body, these tumors can occur in a variety of sites[2]. Although it can be encountered anywhere within the central or periferal nervous system, especially in the neck, thorax, cranium, retroperitoneum, and flexor surfaces of the extremities, localization within the scrotum is extremely rare[3].

In this report, the clinical, radiological and pathological features of a case with

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intrascrotal extratesticular neurofibroma is presented and discussed according to the relevent literature.

CASE REPORT

A 45-year-old man admitted to our clinic with a four year history of gradually enlarging mass and scrotal discomfort in the left scrotum. There was no history of trauma, voiding complaints, or signs and symptoms related to genitourinary disease. On physical examination, a firm, non-tender, mobile, elastic, and nontransilluminating mass was detected in the left hemiscrotum. The size of the lesion was about 3x2 cm and it was seperate from the testis. Systemic examination was unremarkable. All laboratory investigations, including testicular tumor markers, were within normal range. Ultrasonography demonstrated a hypoechoic solitary mass along the margin of the left testis in 3x2x1 cm. Ultrasonographic appearance did not rule out whether this lesion was benign or malign. Abdominopelvic tomography showed a tumoral mass in the left paratesticular region measuring 3x3 cm in size. The other urinary structures were normal. Tomography also showed that, this tumor was seperate from the left testis (Figure 1). Testicular exploration with a preoperative diagnosis of paratesticular tumor was decided and the testis was explored through left scrotal incision. In the operation a solitary mass extending posteriorly toward the internal inguinal ring in 3x3,5 cm diamater was observed (Figure 2). The lesion was dissected from the surrounding tissues and it was noted that the tumor did not involve the other surrounding structures. Frozen examination was performed perioperatively. Frozen evaluation showed a benign proliferative lesion formed by spindle cells and the mass was excised totally. The final histopathologic diagnosis was neurofibroma. Histopathologic examination revealed a tumoral growth which consisted of the fascicles of wavy, spindle Schwannian cells. Focal hypocellular and degenerated areas and vascular structures with thickened wall were observed in the tumor. Immunohistochemically, cytoplasmic S-100 positivity was extensively noted in spindled cells (Figure 3a and 3b). The present case was concordant with neurofibroma on the basis of both light microscopic and immunohistochemical findings. The postoperative period was uneventful with no recurrence after 5 months of follow-up.

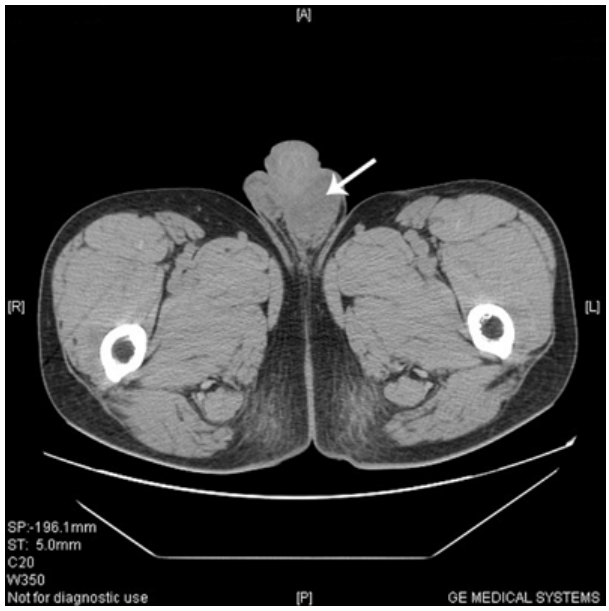

Figure 1: A tumoral mass in the left paratesticular region measuring 3x3 cm in size (arrow) is seen on tomography

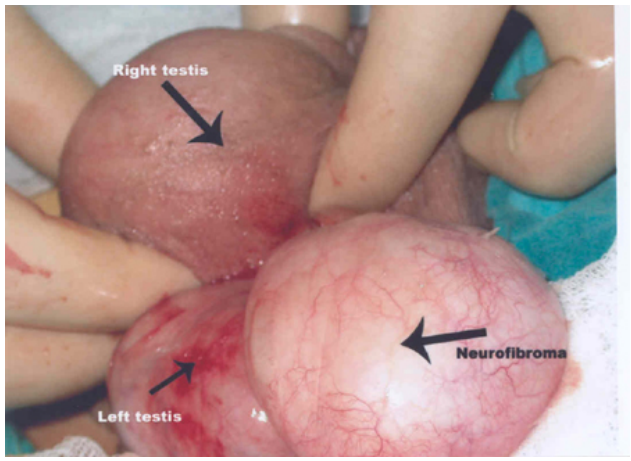

Figure 2: Operation scene of the solitary lesion which has approximately 3x3,5 cm diameters.

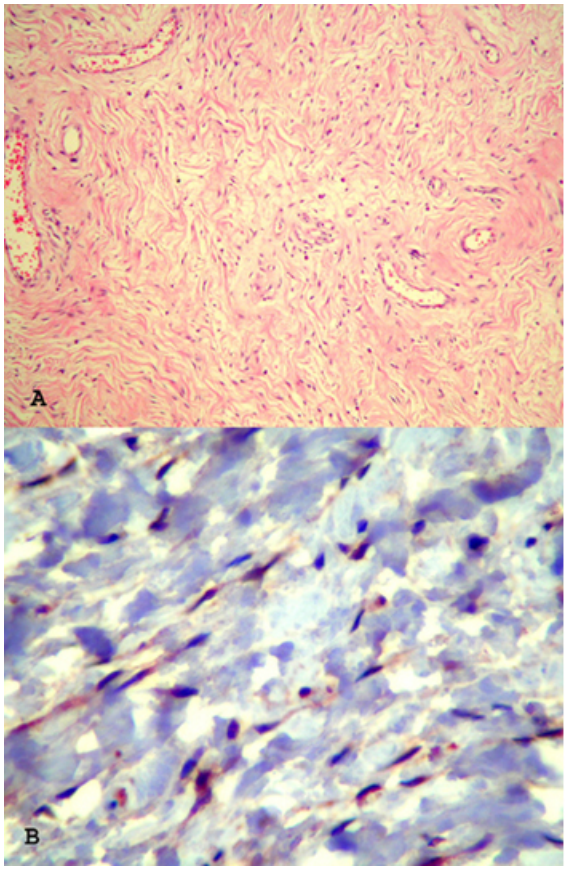

Figure 3a: The spindled and convoluted Schwannian cells in loose, collagenized matrix of the tumor (HE, X25)
Figure 3b. Cytoplasmic S-100 immunostaining of Schwannian cells (AEC, X40)

DISCUSSION

Benign intrascrotal lesions are common findings in the male population[4]. Most of them occur in paratesticular tissue such as epididymis or spermatic cord. Unlike testicular lesions, which are 95% malignant, paratesticular lesions are mostly benign. Usually, surgical exploration is required to rule out intrascrotal malignant processes[4]. Leiomyomas, lipomas, fibromas, hemangiomas and epidermoid cysts have been described as benign tumors of the scrotum. Solitary neurofibroma within the scrotum, which is unassociated with neurofibromatosis is an extremely rare benign tumor[5].

Yamamoto et al., described the first case of solitary neurofibroma in the scrotum[6]. Neurofibroma can be solitary or multiple and presentation age of the cases ranged between 8 and 77 years[1,5]. The presenting complaints are usually scrotal discomfort, painless swelling and hydrocele. The gross appearence of neurofibroma varies a great deal from lesion to lesion[5]. As a rule, the tumors are not encapsulated and have a softer consistency. Microscopically, neurofibromas are formed by combined proliferation of all the elements of peripheral nerve axons, Schwann's cells, fibroblasts, and perineural cells. Schwann's cells are usually the predominant cellular elements in the tumor. These lesions are immunoreactive for S-100 protein and surrounded by basement membrane components as in our case[1,4]. These tumors may originate anatomically from the testis, tunics, and subcutaneous neural tissue. In the literature, eight solitary neurofibroma cases, which involved the external genitalia in the absence of von Recklinghausen's disease, were reported previously[2,5]. In only one case the tumor was localized intratesticularly, whereas in the others the lesion was located extratesticularly[7]. However, the exact origin of the tumor and relation with the intrascrotal components have been somewhat obscure in the reported cases[8,9]. In our patient, we also could not determine the exact anatomic structural origin of the tumor. But we recognized that this mass was separate from the testis, vas deferens, and epididimis. In this case, we agree with Issa et al[10]. that, the tumor must have originated from the genital branch of the genitofemoral nerve lying posteriorly to the spermatic cord. This nerve innervates cremasteric muscle and distributes branches to the skin of the scrotum and adjacent thigh. A similar report has also been published by Milathianakis et al[2].

Although treatment for most primary tumors has historically been radical inguinal orchidectomy, most benign tumors can now be managed by testis sparing surgery[11]. For this reason in neurofibroma, the treatment is surgical excision of tumor[1,3]. In cases, where the tumor involves the testicle, orchidectomy is inevitable. Orchidectomy was performed in only two of the reported cases one because of intratesticular localization of the tumor, the

other because of joint blood supply with the testicle[7,10]. Frozen section microscopic examination should be performed perioperatively to ascertain whether the tumor is benign or malignant[9]. The surgical treatment of testicular and paratesticular tumors is performed by inguinal exploration[11]. But as the mass palpate outside the testis with smooth contours and a four year history of a 3 cm lesion, absence of lymphadenopathies in radiologic evaluations, and normal testis and paratesticular tissues led us to a scrotal surgical approach in this case. Harding et al. reported that 10.4% of men with testicular and paratesticular tumors underwent a scrotal orchidectomy or had a scrotal incision before an inguinal orchidectomy[12]. In their study the authors suggested that scrotal incision is unlikely to affect the risk of loco-regional recurrence. These tumors have an excellent prognosis and they can be cured by complete surgical excision[5,9].

Although an extremely rare disease, intrascrotal extratesticular neurofibroma should be considered in the differential diagnosis of testicular and paratesticular malign tumors.

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