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Mustafa Kemal Üniv Tıp Derg
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OLGU SUNUMU/CASE REPORT

PRIMARY ADRENAL SCHWANNOMA – A RARE CASE

Primer Adrenal Schwannom-Nadir Bir Vaka

Yılmaz Baş[1], İlke Evrim Seçinti[2]

[1]*Hitit Üniversitesi Çorum Eğitim ve Araştırma Hastanesi, Patoloji Bölümü, ÇORUM*
[2]*Sağlık Bakanlığı, Mersin Kamu Hastaneler Birliği Silifke Devlet Hastanesi, Patoloji Laboratuvarı, MERSİN*

ABSTRACT

Solitary-cystic Schwannoma arising from adrenal gland is seen extremely rare. Its frequency in our country is unknown. Schwannomas are responsible for 1-5% of all retroperitoneal masses and clinical signs are non-specific. They are diagnosed incidentally as in our case and generally their first sign is referred pain. CT and MR are important screening methods. However, the final diagnosis is made by pathological examination. The prognosis is excellent after total resection. It is valuable to present the primary adrenal schwannoma arising from adrenal gland because of its rare incidence in all of the world thus this is the first case in our country.

Key words: Adrenal gland, schwann cells, neoplasm.

ÖZET

Adrenal bezden köken alan soliter kistik schwannom oldukça nadirdir. Ülkemizdeki görülme sıklığı bilinmemektedir. Retroperitoneal kitlelerin %1-5'ini oluşturmaktadır. Klinik bulguları non-spesifiktir. Bizim olgumuzda olduğu gibi tesadüfen saptanırlar ve genel olarak ilk bulguları ağrıdır. Tanıda BT ve MR önemli görüntüleme yöntemleridir ancak kesin tanı patolojik incelemeyle konulur. Rezeksiyondan sonra prognoz mükemmeldir. Adrenal bezden köken alan primer adrenal schwannom dünyada nadir olarak saptanması ve ülkemizde ilk vaka olması nedeniyle yayınlanmaya değer görülmüştür.

Anahtar kelimeler: Adrenal bez, schwann hücreleri, neoplazm.

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İletişim: İlke Evrim Seçinti Sağlık Bakanlığı, Mersin Kamu Hastaneler Birliği, Silifke Devlet Hastanesi, Patoloji Laboratuvarı, Silifke, MERSİN/Türkiye
Tel: 0 5066899626 **E-posta:** ilkevrim@gmail.com

INTRODUCTION

Solitary-cystic Schwannoma arising from adrenal gland is seen extremely rare. Its frequency in our country is unknown. Schwannomas are responsible for 1-5% of all retroperitoneal masses (1). Its clinical signs are non-specific. They are diagnosed incidentally as in our case and generally their first sign is referred pain. CT and MR are important screening methods. However, the final diagnosis is made by pathological examination. The prognosis is excellent after total resection. It is valuable to present the primary adrenal schwannoma arising from adrenal gland because of its rare incidence in all of the world thus this is the first case in our country.

CASE REPORT

63-years old female patient admitted our hospital for the pain of right upper quadrant. Her physical examination, blood pressure and blood and urine biochemistry were in normal limits, including serum cortisol and urine vanilmandelic acid (VMA) and methanephrin. Well-circumscribed mass in right adrenal gland (Figure 1) was detected on ultrasonography and computed tomography. Encapsulated, lobulated, dirty-white-cream colored and elastic mass, in appearance, was resected totally (Figure2). The surface of section has nodular and fibrous appearing and solid structure at areas close to capsule (Figure 2). In the areas close to middle, there were some cystic openings. Tumor consisted of short-crossed fascicles and benign fusiform cells which were showing nuclear “palisading” and curved path in some areas (Figure 3). Pleomorphism, hyperchromasia, marked nucleus, mitosis and necrosis were not found. Hyaline thickening in vascular structures was remarkable. Whole mass was examined but histomorphological signs belong to normal adrenal tissue was not determined.

In the immunohistochemical examination, tumor cells were stained positive for S-100 (Figure 4), Synaptophysin (Figure 5) and Neuron Specific Enolase (NSE) (Figure 6) while it was not observed in staining for SMA (Actin).

Figure 1. On Computed Tomography was determined a well-circumscribed mass in right adrenal gland (Arrow).

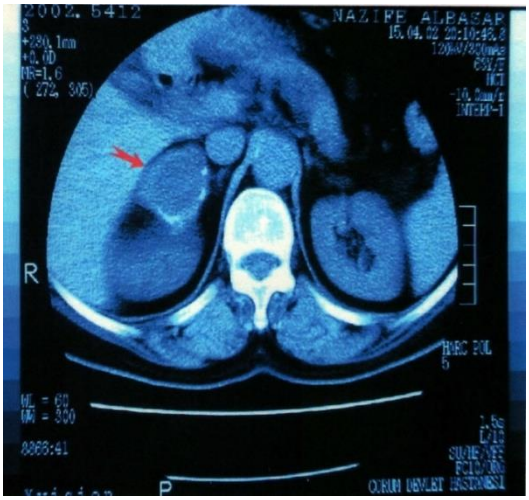


Figure 2. The surface of section has nodular and fibrous appearing and solid structure at areas close to capsule.

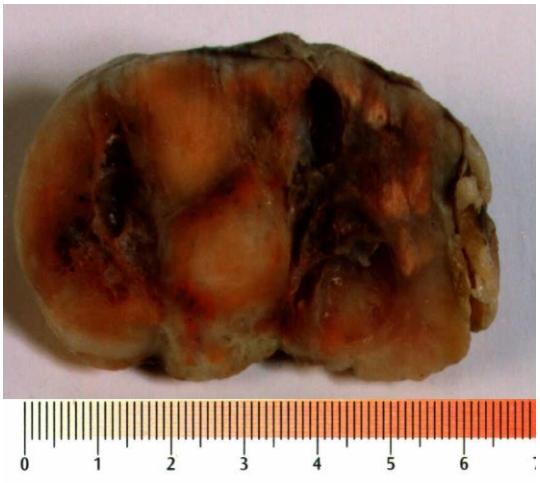


Figure 3. Tumor consisted of short crossed fascicles and benign fusiform cells making from place to place nuclear "palisading" and having curved path(H.Ex 200).

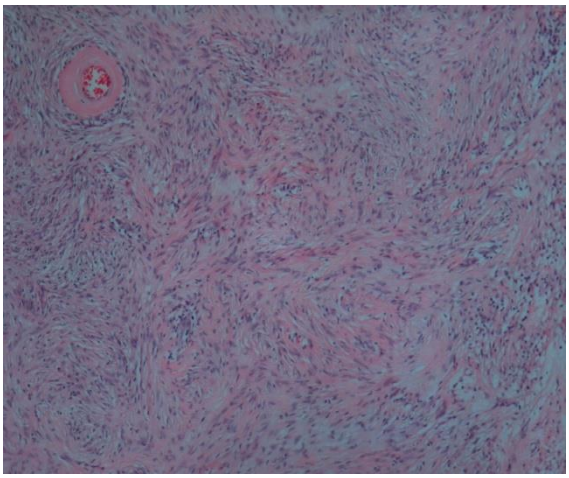


Figure 4. Immunostain for S100 (x400)

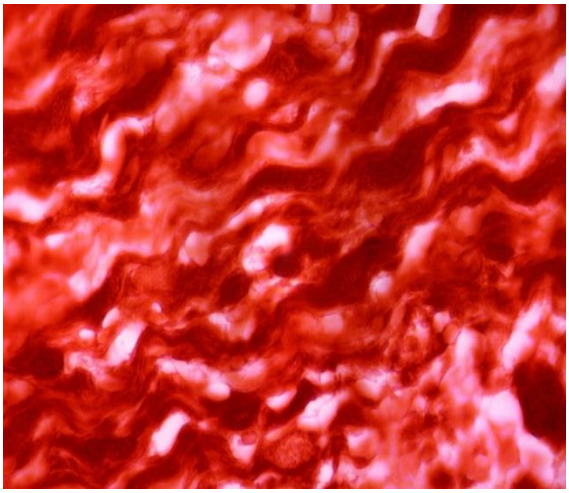


Figure 5. Immunostain for Synapthofyzin (x 400).

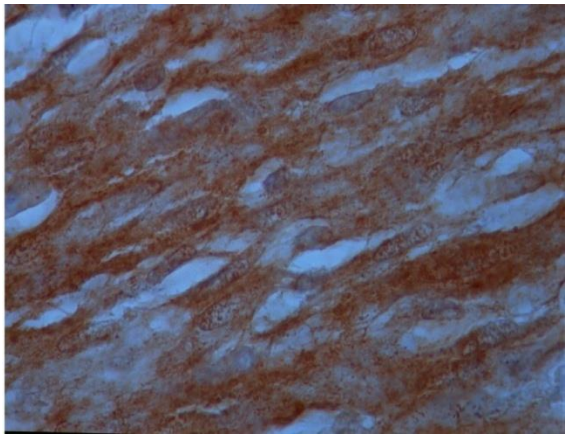
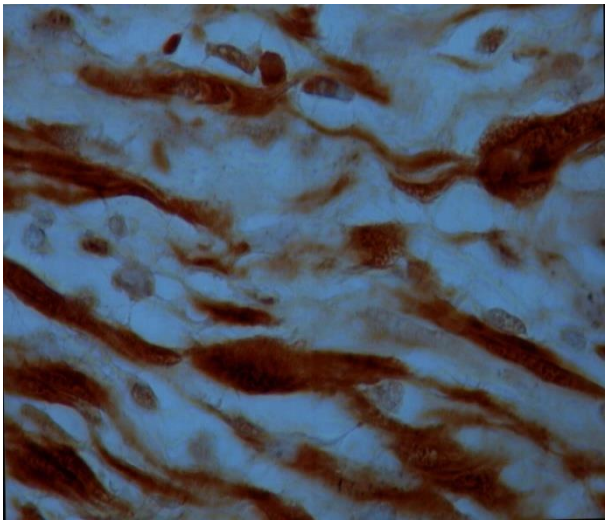


Figure 6. Immunostain for NSE (x 400)



DISCUSSION

Neural system-derived adrenal gland tumors involve primary orjuxta-adrenal schwannoma, adrenal neurofibroma and primary adrenal malignant schwannoma (1,2,3,4).

Schwannomas develop from Schwann cells wich are originating from neural crest (1,4,5,6,7). The most common Schwannomas occur as slow-growing and benign nerves heath tumors in head, neck, upper and lower extremities and the trunk. Schwannomas were foundonly 1% in a large series including 688 retroperitoneal tumors (2) and constitute 1-10% of all retroperitoneal masses (6,7). Retroperitoneal Schwannomas are usually presented with abdominal or back pain but the symptoms are variable (4,6). Preoperative diagnosis is difficult. Most of these cases detected incidentally. The diagnosis has been always made after pathological examination (4,6,8).

The cystic openings occur depending on parenchymal degeneration in nonfunctional adrenal Schwannomas which have not significant clinicale vidence for a long time before diagnosis. The presence of cystic

changes together with a retroperitoneal tumor are frequently seen and thise vidence guide to diagnosis of nonfunctional adrenal schwannoma (6, 9).

Benign schwannomas are solitary, homogenous, well-defined and encapsulated masses and do not invade the neighbouring organs and tissues (5). Like the present case, benign schwannomas comprise of Antoni A area which have cystic structures and fusiform cells showing palisadic design, and hypocellular Antony B area. There were not typicaland/or atypical mitosis, pleomorphism, hyperchromasia, prominent nucleolus, necrosis and capsule and/or angiolymphatic invasion in our case.

The present case is valuable for presentation because it has been revealed that an adrenal-derived mass should be considered if there is not any ethiological reason in the patients who is admitted the clinic for abdominal and/or back pain. When reviewed the literature, it was found that Primary Adrenal Schwannoma was not observed in our country so far and the present case would be acceptable as the first case.

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