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Primary Paratesticular Ganglioneuroma: A Rare Case in an Adult

Primer Paratestiküler Ganglionörom: Erişkinde Nadir Bir Olgu

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ABSTRACT

Ganglioneuroma is a benign tumor, affecting the central nervous system, commonly seen in children and young adults. Non-cranial ganglioneuromas are seen predominantly in the posterior mediastinum and retroperitoneum. Paratesticular neoplasms are extremely rare and are susceptible to clinical misinterpretation as malignancy. Tumors of nerve origin are a rarity in the paratesticular region, with a few cases of schwannoma and malignant peripheral nerve sheath tumor being reviewed in the literature. A rare case of ganglioneuroma involving the paratesticular region in an adult is presented.

Key Words: Paratesticular, Ganglioneuroma, Adult

ÖZ

Gangliyonöroma genellikle çocuklarda ve genç erişkinlerde görülen benign bir santral sinir sistemi tümörüdür. Kraniyum dışındaki gangliyonöromlar posterior mediasten ve retroperitoneal bölgede görülürler. Paratestiküler tümörler son derece nadirdir ve yanlışlıkla klinik olarak malign tümör şeklinde değerlendirilebilir. Schwannom ve malign periferik sinir kılıfı tümörü olarak literatürde gözden geçirilmiş az sayıda olgu mevcut olup bu bölgenin sinir kökenli tümörleri seyrektir. Yetişkinde paratestiküler bölgede nadir görülen ganglionörom olgusu sunulmaktadır.

Anahtar Sözcükler: Paratestiküler, Ganglionörom, Erişkin

INTRODUCTION

Neuroblastoma, ganglioneuroblastoma, and ganglioneuroma are tumors of the sympathetic nervous system that arise from primitive sympathogonia. The most benign tumor is the ganglioneuroma, which is composed of gangliocytes and mature stroma. It is usually seen in children and young adults and most often involves the central nervous system (1). Most non-cranial ganglioneuromas are located in the posterior mediastinum and the retroperitoneum (1). Involvement of the paratesticular region, as in this case, is a rarity, with the patient undergoing surgical intervention for diagnostic and therapeutic purposes.

CASE REPORT

A 23-year-old male presented with painless left scrotal swelling of one year duration. The swelling was insidious in onset and gradually progressive. On examination, the swelling was smooth and mobile, measuring 7x6x3 cm in size, soft to firm in consistency, with transillumination being positive. A provisional clinical diagnosis of a benign tumor - lipoma of the epididymis was made. Scrotal exploration was undertaken and the paratesticular mass was excised with preservation of the testis.

On gross examination, an encapsulated lesion measuring 7x6x3 cm was identified (Figure 1A). The cut section of the lesion revealed homogenous grey white areas with focal whorling and myxoid regions with absence of hemorrhage and necrosis (Figure 1B). Histopathologically, an encapsulated lesion composed of few large round to oval cells (Figure 2) with abundant amphophilic cytoplasm, cytoplasmic basophilic granules, large eccentric vesicular nucleus with a prominent nucleolus (Figure 3), along with few binucleate ganglion cells, and loosely arranged spindle cells with wavy, buckled nuclei in a fibrillary matrix was noted. Based on these features, the diagnosis was ganglioneuroma, maturing type.

DISCUSSION

Neuroblastoma, ganglioneuroblastoma, and ganglioneuroma are tumors of the sympathetic nervous system originating from primitive sympathogonia and are referred to collectively as neuroblastic tumors. They arise wherever sympathetic tissue exists and may be seen in the neck, posterior mediastinum, adrenal gland, retroperitoneum, and pelvis (1). The three tumors differ in their degree of cellular and extracellular maturation. Immature tumors tend to be aggressive and occur in younger patients

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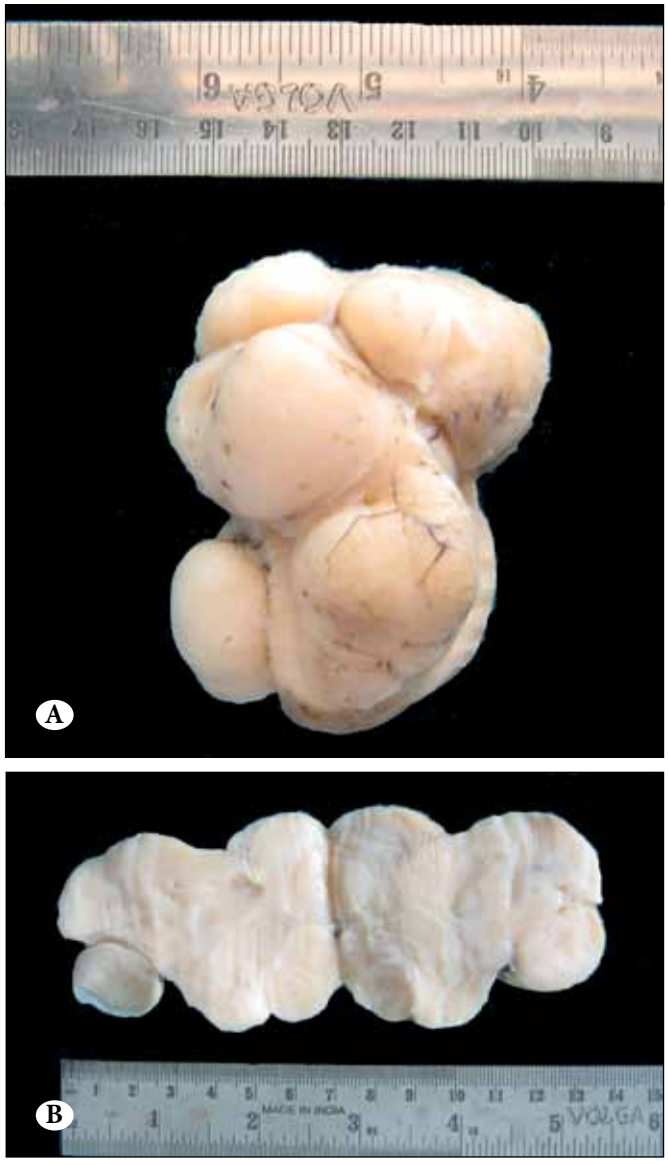

Figure 1: Photograph showing an encapsulated lesion **(A)** and the cut section showing homogenous grey white areas with focal whorling and myxoid regions **(B)**.

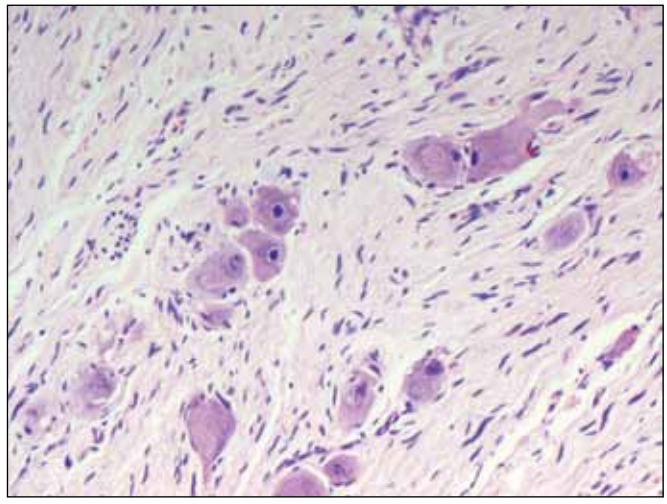

Figure 2: Nodules of ganglion cells in various stages of maturation against a background of neurofibroma-like stroma. (H&E, x100).

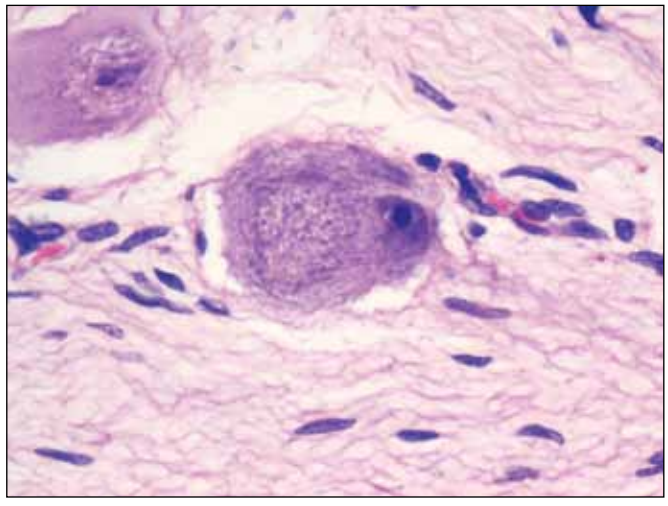

Figure 3: Two ganglion cells with abundant eosinophilic cytoplasm, intracytoplasmic Nissl granules, and eccentric ovoid nucleus with prominent eosinophilic nucleolus. (H&E, x400).

(median age, under 2 years), whereas mature tumors occur in older children (median age, approximately 7 years) and tend to behave in a benign fashion. Ganglioneuromas, neuroblastomas, paragangliomas, and pheochromocytomas are thought to have close embryogenic, structural, and functional similarity. These are regarded as neural crest neoplasms with close anatomic relations to blood vessels. The most benign tumor is the ganglioneuroma, which is composed of neurites, Schwann cells and relatively mature ganglion cells. Most ganglioneuromas seem to arise de novo; however, maturation of neuroblastoma to ganglioneuroma has been documented. This tumor is usually seen in the 10- to 30-year age group with an adult onset as in our case being extremely rare. Microscopically, ganglioneuromas are designated as mature and maturing subtypes, based on whether every single ganglion cell is mature, or there is a minor component of scattered collections of differentiating neuroblasts and/or maturing ganglion cells. This case histopathologically showed maturing ganglion and was subtyped likewise.

Noncranial ganglioneuromas are most often located along the paraspinal structures in the posterior mediastinum or retroperitoneum and are capable of metastasizing in a small percentage of cases (1). Therefore, surgical removal is considered essential for a more favorable prognosis. Pathological evaluation is essential for the establishment of the diagnosis and to obviate the development of malignancy

or a focus of neuroblastoma (1). Most authors also suggest magnetic resonance imaging to rule out metastasis in the rest of the body (1,6).

The paratesticular region consists of many cell types and neoplasms arising therefrom are histologically heterogeneous. Tumors in this region present as discrete scrotal masses and may cause diagnostic confusion with testicular tumors. The most common benign tumors are lipomata, adenomatoid tumors and leiomyomata; the more common malignant neoplasms are sarcomas, mesotheliomas, lymphomas, epididymal adenocarcinomas and rarely metastases (2,3). Ganglioneuromas involving the paratesticular structures, as in our case, are extremely rare, with the first such case reported by Ceroni and Di Luttichau in a child (4). A search in the English electronic media yields only four reported cases of paratesticular ganglioneuromas, two each in children and adults including one reported by Banks et al., in a 15-year-old boy diagnosed as composite paratesticular neoplasm consisting of mixed ganglioneuroma and malignant peripheral nerve sheath tumor features (4,5,6).

Ganglioneuroma, an otherwise benign tumor, appears to have the rare capacity to develop into neuroblastoma in a delayed fashion. This is substantiated by a documented case of spinal neuroblastoma in a 32-year-old woman, who had presented with a history of retroperitoneal ganglioneuroma 11 years earlier. The patient has survived 10 more years with relatively stable disease and the recent development of metastases. This is either because of dedifferentiation or the long-term presence of a unique, quiescent form of neuroblastoma (7). This makes early detection with surgical excision necessary to prevent the remote occurrence of malignancy at a later date.

Paratesticular ganglioneuroma is a benign and rarely occurring tumor that can often lead to clinical misinterpretation and unnecessary mutilating surgeries. The surgeon would do well to keep this rare tumor in mind when dealing with benign paratesticular lesions in a young adult or child. Owing to the rare propensity to develop into malignancy and to prevent a risk of dedifferentiation, excision of the lesion is the best treatment modality available.

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