

# Adrenal ganglioneuroma with osteoid metaplasia: report of a case

Nese Ekinici<sup>1</sup>, Seyran Yigit<sup>1</sup>, Mustafa Ozer<sup>2</sup>

<sup>1</sup>Sonuç Medical laboratory, <sup>2</sup>Private Hayat Hospital, Izmir, Turkey

Accepted for publication on 27 July 2004

We report herein an unusual case of ganglioneuroma of the adrenal gland with osteoid metaplasia. A 46-year-old man was admitted to the hospital with a left adrenal mass of 10 cm in diameter found incidentally. The mass was nonfunctioning. Surgery was performed including left adrenalectomy and resection of an additional mass in the surrounding adipose tissue with a diameter of 1.5 cm. Histopathological examination revealed the diagnosis of ganglioneuroma for both lesions. Osteoid metaplasia was found in the adrenal mass that has not been reported before. We conclude that adrenal ganglioneuromas should be kept in mind in the differential diagnosis of adrenal gland lesions found incidentally.

**Key words:** Adrenal, ganglioneuroma, osteoid, metaplasia

## Introduction

Ganglioneuromas are benign and differentiated tumors of the primordial neural crest cells. They are mostly located in the posterior mediastinum and retroperitoneum. Their location in the adrenal glands are less common but when occurs, it represents a diagnostic challenge. Characteristically they do not secrete catecholamines or steroid hormones, hence were regarded as silent lesions and found usually incidentally. Because its rarity, it may not included in the differential diagnosis of adrenal lesions.

Herein, we report a very rare case of multiple ganglioneuroma located in the left adrenal gland and surrounding adipose tissue. Osteoid metaplasia was found in the adrenal tumor, and to the best of our knowledge, this metaplasia has not been reported before.

## Case

A 46-year-old man who had been under treatment for lung tuberculosis for two years underwent routine abdominal ultrasonography followed by computed

tomography which revealed a large lesion (10 x 8.0 x 8.0 cm) originating from the left adrenal gland. The lesion was well circumscribed from the surrounding tissue and had a fine granular calcification without any evidence of necrosis, hemorrhage, local extension or invasion. The patient's history did not represent any systemic symptoms. Laboratory results were within normal limits except for an increased sedimentation. Bblood levels of the adrenal hormones and their urinary metabolites were normal

He underwent an elective operation for the adrenal mass. The mass had smooth borders, was soft in consistency and located inside of the adrenal gland. Another mass with a diameter of 1.5 cm was observed in the neighbouring adipose tissue. The lesions were totally resected in an en-block manner including left adrenal gland and peripheral adipose tissue.

Macroscopic examination of the resected specimen in the adrenal revealed an encapsulated tumor of 400 g in weight and 10x 8x 8 cm in diameter. The cut surface was solid, soft to firm, somewhat fibrous and whorled, gray-tan, partially brown in color. Adjacent to the main tumor mass, a second tumor of 2

g in weight and 1.2 cm in diameter was detected in the periadrenal fat tissue.

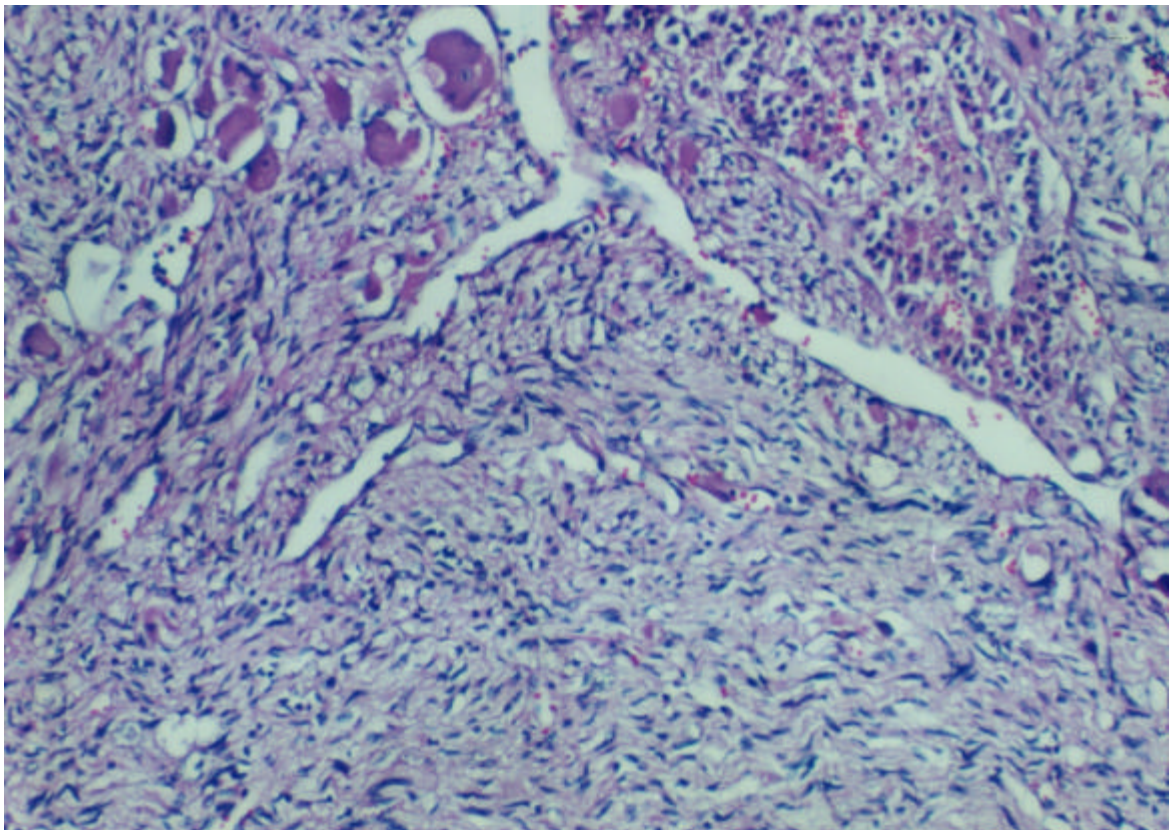
Microscopic examination showed that both tumors consisted of bundles of Schwann cells in the background which crisscross each other in a longitudinal and transverse fashion. Numerous mature ganglion cells either isolated or in small clusters were scattered throughout the tumor (Figure 1). Fontana-positive pigment in some of the stromal and ganglion cells, coarse patchy calcification, adipose tissue in the stroma, cystic change and focal lymphocytic infiltrate were also found. Detailed examination showed no immature elements in both tumors. The histopathological characteristics showed that both tumors were ganglioneuroma. An additional finding was the presence of osteoid metaplasia in the fat tissue of the larger tumor's stroma (Figure 2).

The patient was discharged from the hospital on the 3rd postoperative day and has been well for six months after the surgery.

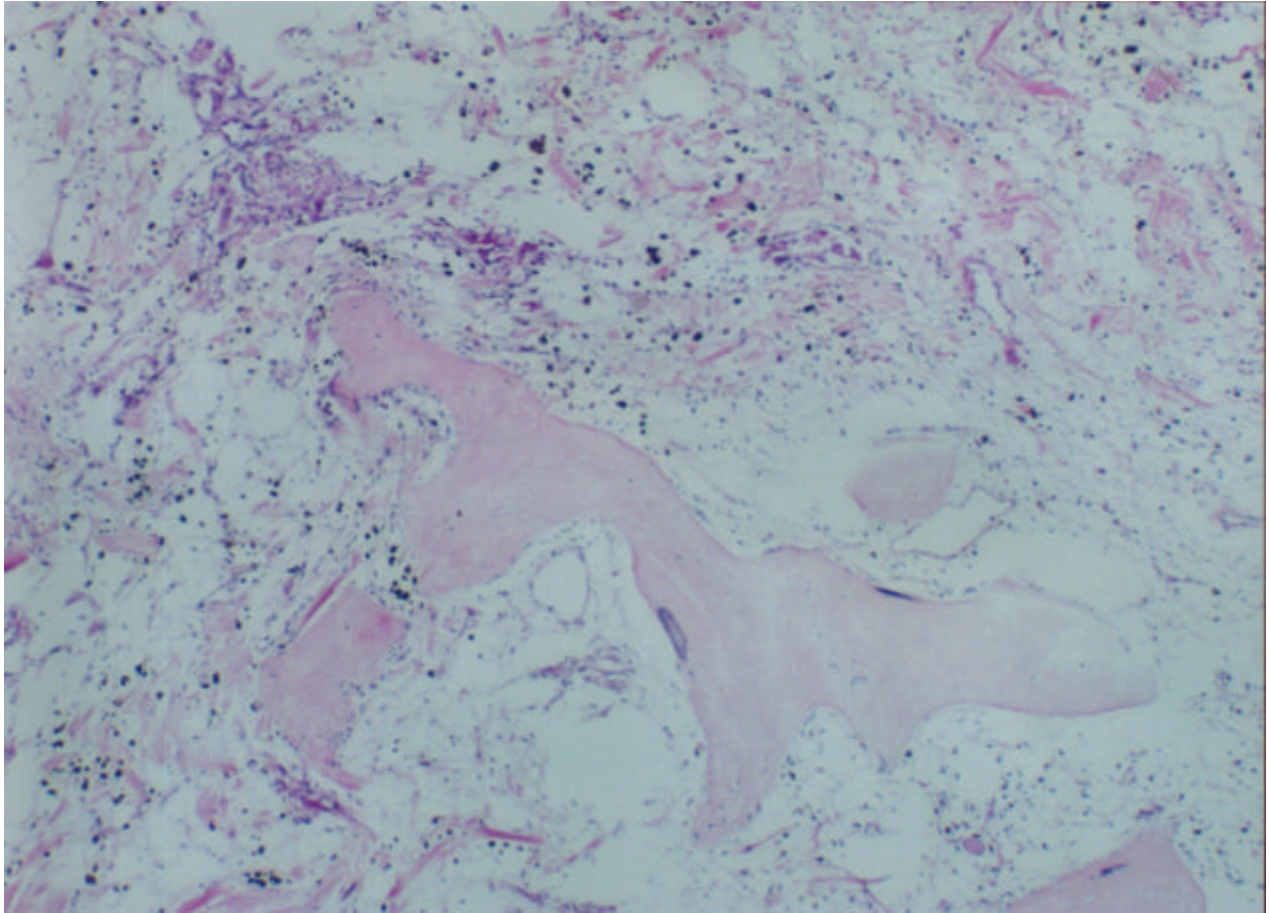
## Discussion

Ganglioneuroma, a benign tumor of the sympathetic nervous system may arise anywhere along the paravertebral sympathetic plexus and occasionally originate from the medulla in the adrenal gland (1, 2). They are mostly located in the posterior mediastinum, retroperitoneum, pelvis and adrenal medulla. However, unusual localizations including heart, gastrointestinal tract, cervix and skin have been also reported (1-4). Its central nervous system counterpart is called ganglioglioma. Ganglioneuromas occur in all age groups, but are often diagnosed in patients older than 10 years (1, 2). Most adrenal ganglioneuromas do not secrete exogenous hormones and thus the patients usually have a clinically silent course (1, 2, 5). The tumor is often found incidentally in the patients undergoing radiological investigations for some other reasons as seen in our patient (5).

Although nearly all ganglioneuromas are solitary, multiple lesions have been rarely reported (1). Our



**Figure 1.** Scattered ganglion cells in the background of Schwann cells, H+E x 110.



**Figure 2.** Osteoid metaplasia of fat tissue in the tumor stroma, H-E x 220.

case is unique on this respect for the reason that the additional lesion, although not identified by preoperative imaging tools, was found adjacent to the larger mass with a careful exploration of the surgical area.

Ganglioneuromas are composed of fully differentiated elements resembling mature autonomic ganglion cells and satellite cells in a fibrous stroma with an abundance of unmyelinated as well as occasional myelinated axons of Schwann cells. Degenerative changes are frequently detected in ganglioneuromas. These include accumulation of brown cytoplasmic melanin like substance (neuromelanin) believed to represent catecholamine products after autooxidation and also stromal degenerative changes particularly in large tumors (1, 2). Fibrosis and some degrees of mucin accumulation, occasional adipose tissue infiltration, cystic change, lymphocytic infiltrate, coarse patchy calcification and rare psammoma bodies may be seen. To the best of our knowledge, osteoid metaplasia of the fat tissue in the

stroma of ganglioneuroma found in the present case has not been reported before.

## References

1. Scheithauer BW, Woodruff JM, Erlandson RA: Benign neurogenic tumors, in Rosai J, Sobin LH (eds). *Atlas of Tumor Pathology, Tumors of the Peripheral Nervous System*. Armed Forces Institute of Pathology. Washington DC, 1999, pp 259–276.
2. Weiss SW, Goldblum J: Primitive neuroectodermal tumors and related lesions, in: Strauss M (ed). *Enzinger and Weiss's Soft Tissue Tumors*. Mosby, Inc, St Louis, Missouri, 4th ed., 2001, pp: 1284–1288.
3. Çelik V, Ünal G, Özgültekin R, Göksel S, Ünal H, Çerçel A. Adrenal ganglioneuroma. *Br J Surg* 1996; 83:263.
4. Hammond RR, Walton JC. Cutaneous ganglioneuromas : A case report and review of the literature . *Hum Pathol* 1996;27:735–738.
5. Johnson GL, Hruban RH, Marshall FF, Fishman EK. Primary adrenal ganglioneuroma : CT findings in four patients. *Am J Radiol* 1997; 169: 169–171.