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SHORT REPORT

Giant Juxtadrenal and Adrenal Schwannoma with Concurrent Adrenal Myelolipoma Mimicking an Adrenal Malignant Tumor*

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Schwannomas are common benign tumors, composed of differentiated neoplastic Schwann cells. The majority arise from peripheral nerves in the head and neck region and nerves of the upper extremity (1). Visceral schwannomas, in particular, juxtadrenal and adrenal schwannomas are exceedingly rare (2,3,4). Adrenal myelolipomas are composed of mature adipose and hematopoetic elements that resemble bone marrow. It was first described by Grieke in 1905, and was named "myelolipoma" by Oberling in 1929 (5,6). Here we describe a giant juxtadrenal and adrenal schwannoma with concurrent adrenal myelolipoma mimicking an adrenal malignant tumor.

Case Report

A 46 -year-old woman was referred to our hospital for further evaluation and treatment of a left adrenal mass. The patient was initially admitted to a local clinic with complaints of a menstrual disorder. During investigations, ultrasound and CT scanning revealed a 8x8 cm mass arising from the region of the left adrenal. The size of the tumor was suggestive of an adrenal malignant tumor. The patient was referred for adrenalectomy and complete resection of the retroperitoneal mass. Serum electrolytes, complete blood count, aldosterone, cortisol, and urine catecholamines were within normal limits, except the serum calcium level, which was at the upper limit (10.5mg/dl). Total excision of the mass with left adrenelectomy was performed.

The resected specimen weighed 254g and measured 9.5x8.5x8 cm in size. A tumor measuring 8x7.5x6cm was present in the periadrenal fat tissue just next to the left adrenal gland. The adrenal gland was 6x2.2x1.3cm in size. The tumor was located in the vicinity of, but not connected to the adrenal gland (Figure 1A). The tumor was encapsulated, solid, and yellow-white in color. Hemorrhage or necrosis was not noticed. On the other hand, another mass was noticed in the adrenal gland, which was 1.8x1.3x0.8 cm in size, solid, white, and relatively hard in consistency (Figure 1B).

Histologically, the juxtadrenal tumor was completely encapsulated with fibrous tissue. The tumor was cellular and composed of spindle cells arranged in a fascicular pattern (Figure 1C). There was perivascular hyalinization and peritumoral lymphocytic infiltration. Necrosis and mitoses were not observed. Immunohistochemically, the tumor cells were positive for S-100 protein (Figure 1D) whereas SMA, CD34, Desmin, EMA, and NFP were negative.

* This case was presented at XXIII. World Congress of Pathology and Laboratory Medicine in poster section.

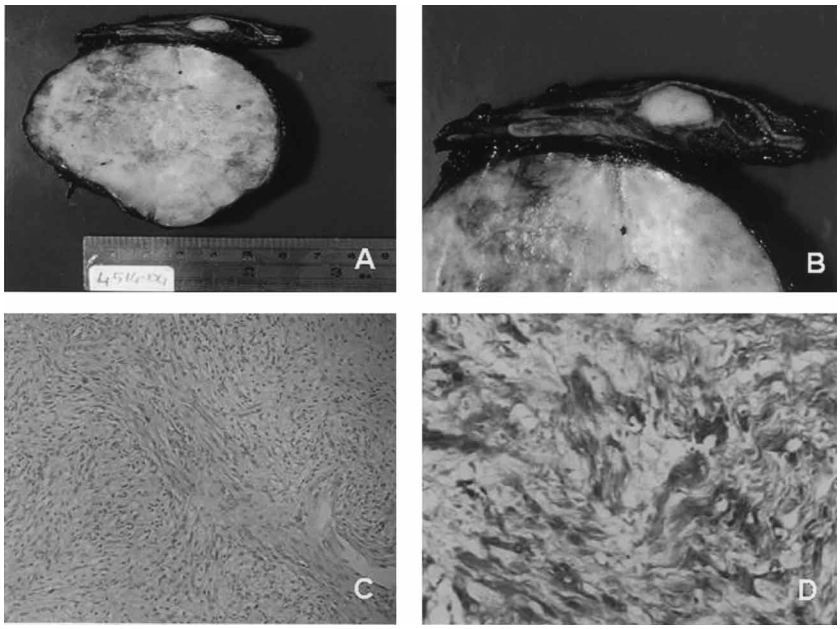

Figure 1. Gross photograph of a adrenal and juxtadrenal tumor. The specimen consists of cut sections of a adrenal gland (upper) and a juxtadrenal mass (A,B). The microscopic section of juxtadrenal schwannoma. The tumor is composed of spindle cells arranged in a fascicular pattern (C) (HEX100). Intense cytoplasmic staining with S-100 of juxtadrenal mass (D) (IHCX200).

The microscopic examination of the left adrenelectomy specimen showed normal adrenal histology peripherally but centrally a mixture of spindle cell tumor and myelolipoma characterized by mature adipocytes and hematopoetic cells were recognized (Figure 2). The spindle cell tumor had similar morphological features to the juxtadrenal tumor. The adrenal tumor was less cellular and contained few mesenchymal spindle cells. No mitosis was observed. There was only minimal lymphoreticular reaction in the surrounding adrenal parenchyma. Numerous serial sections were taken and it was determined that there was no connection between the juxtadrenal and adrenal tumors. Immunohistochemical studies revealed that S-100 was also strongly positive whereas SMA, CD34, Desmin, EMA, and NFP were negative in the adrenal neoplastic cells. The diagnosis of juxtadrenal and adrenal schwannoma with concurrent adrenal myelolipoma was made.

To verify high calcium level, parathormone (PTH) level was examined and was found to be 105pg/dl. Parathyroid Tc^{99m}-MIBI scanning revealed parathyroid adenoma and parathyroidectomy was performed 2 months after adrenelectomy. Histologically, the diagnosis of parathyroid adenoma was made. Postoperatively, the patient's serum calcium and parathormone levels returned to normal. Clinically, there was no history of neurofibromatosis type 2 and multiple endocrine neoplasia type 1. Currently, the patient is under follow-up with no recurrence one year after the initial operation.

Schwannomas are usually benign tumors that arise from the Schwann cell (1). Although they can occur wherever Schwann cells are present, visceral schwannomas, particularly juxtadrenal and adrenal schwannomas, are exceedingly rare; only a few cases have been reported in the English literature (2,3,4). Most of them were preoperatively diagnosed as adrenal tumors. In our case, the size of the tumor was suggestive of an adrenal malignant tumor preoperatively. Adrenal myelolipomas are usually asymptomatic and relatively rare lesions composed of mature adipose tissue and hematopoetic elements. They are found incidentally at radiological examination or autopsy (5,6,7). To our knowledge, the presence of concommitant myelolipoma within adrenal tissue has not previously been reported in patients with adrenal-juxtadrenal schwannomas.

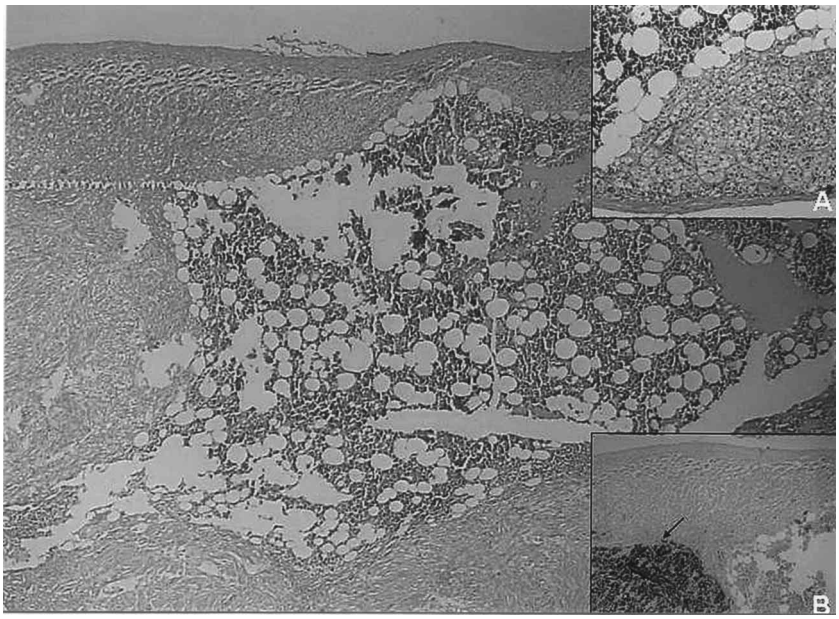

Figure 2. Mixture of spindle cell tumor and myelolipoma surrounded by normal adrenocortical tissue. Myelolipoma composed of normal-appearing hematopoietic cells and adipocytes with adjacent normal adrenocortical tissue visible at the top inset (A). Positive immunoreactivity of spindle cell tumor (arrow) for S-100 antibody visible at the bottom inset (B).

Although most cases represent incidental findings in asymptomatic patients, adrenal myelolipomas arising in patients with endocrine abnormalities such as Cushing syndrome and congenital adrenal hyperplasia have been reported (8,9). Since pathogenesis of myelolipomas is poorly understood, it is unclear in this case whether this occurrence is merely incidental or somehow related to the presence of schwannomas and parathyroid adenoma. Schwannomas predominantly arise from peripheral nerves in the head and neck region and the upper extremity. Histopathologically schwannomas have characteristic morphologic patterns. They are benign tumors, but the malignant and unusual forms have also been described. To distinguish schwannoma from malignant peripheral nerve sheath tumor and leiomyosarcoma, the number of mitotic figures and necrosis are the most important cellular features. Bilateral vestibular schwannomas are pathognomonic of neurofibromatosis type 2 (NF2), while multiple peripheral schwannomas in the absence of other NF2 features is characteristic of schwannomatosis, a newly described syndrome (10). In summary, schwannomas are very rare benign tumors of the juxtadrenal –adrenal gland and may easily be confused with adrenal malignant tumor. Although very rare, juxtadrenal-adrenal schwannomas should be considered in the differential diagnosis of any retroperitoneal mass. We report, to our knowledge, the first case of giant juxtadrenal and adrenal schwannomas presenting as an adrenal malignant tumor with a simultaneously occuring myelolipoma in the same adrenal and adenoma of the parathyroid gland.

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