Title	Solitary retroperitoneal neurofibroma: a case report
Author(s)	ISHIKAWA, Jiro; KAMIDONO, Sadao; MAEDA, Sakan; SUGIYAMA, Taketoshi; HARA, Isao; TAKECHI, Yoshizumi; OHMAE, Hiroshi; HARA, Shinji
Citation	泌尿器科紀要 (1989), 35(7): 1157-1160
Issue Date	1989-07
URL	http://hdl.handle.net/2433/116604
Right	
Type	Departmental Bulletin Paper
Textversion	publisher

Acta Urol. Jpn. 35: 1157-1160, 1989

SOLITARY RETROPERITONEAL NEUROFIBROMA: A CASE REPORT

Jiro ISHIKAWA and Sadao KAMIDONO

From the Department of Urology, Kobe University School of Medicine

Sakan MAEDA and Taketoshi SUGIYAMA

From the Department of Pathology, Kobe University School of Medicine

Isao HARA, Yoshizumi TAKECHI, Hiroshi OHMAE and Shinji HARA

From Hara Genitourinary Hospital

A solitary neurofibroma arising in the retroperitoneal space without any other stagma of von Recklinghausen's disease is reported. Confusion with another nerve sheath tumor, a schwannoma is a diagnostic pitfall. Histochemical and immunohistochemical stainings of the tumor are useful for the diagnosis of solitary neurofibroma.

Key words: Solitary neurofibroma, Retroperitoneal space, Alcian blue staining, S-100, NSE

INTRODUCTION

A neurofibroma is a benign nerve sheath tumor that occurs as a solitary tumor or as a partial manifestation of von Recklinghausen's disease : an inheritant disease characterized by multiple cutaneous pigmented lesions (cafe-au-lait spots) and subcutaneous neurofibromas. Although genitourinary involvement of neurofibromas is rare, such cases have been reported in the bladder, kidney, ureter, penis, and retroperitoneal space[1-6]. Previous literature shows confusion relating to the terminology of the peripheral nerve tumors[7]. We report a case of a solitary retroperitoneal neurofibroma diagnosed by histochemical and immunohistochemical stainings as well as light microscopic appearance of the tumor.

CASE REPORT

A 56-year-old Japanese woman presented with a 2-year history of dysuria which fluctuated with periods of exacerbations and remissions. Significant medical history included total abdominal hysterectomy for leiomyomas 22 years earlier. Family history was negative for von Recklinghausen's disease. Physical examination revealed traction pain in the left lower limb. Cutaneous examination revealed no cafe-au-lait spots or neurofibromas. All the hematological and serum biochemical data, and urinalysis revealed no abnormality. Drip infusion pyelogram (DIP) demonstrated the medially dislocated bladder and the left ureter, suggesting the existence of the extravesical mass (Fig. 1). On cystoscopy, the bladder mucosa was intact. Computerized tomography (CT) demonstrated a well encapsulated homogeneous mass in the retrovesical space (Fig. 2). Exploratory laparotomy, performed on May 2 1987, revealed an elastic soft tumor in the retrovesical space. The $8 \times 8.5 \times 8$ cm tumor, weighing 326 g was completely excised. Cut surface of the tumor was yellowish-gray in color, and gelatinous. Microscopic examination of the excised tumor revealed fusiform tumor cells in numerous collagen fibrils and a non-oraganized matrix (Fig. 3). The cell arrangement of Antoni type A and type B was not observed. No cellular atypism and mitotic figures were observed. Alcain blue staining showed positive findings in the matrix. Immunohistostaining for S-100 protein and neuron specific enolase (NSE) showed positive in the tumor cells

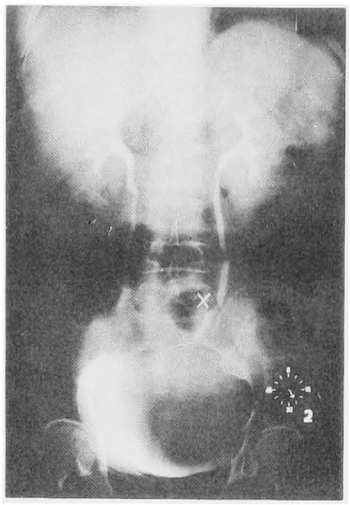

Fig. 1. DIP shows the displacement of the bladder and left ureter medially, suggesting a large extravesical mass.

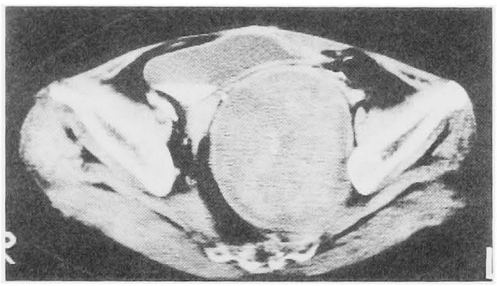

Fig. 2. CT demonstrates a retrovesical mass.

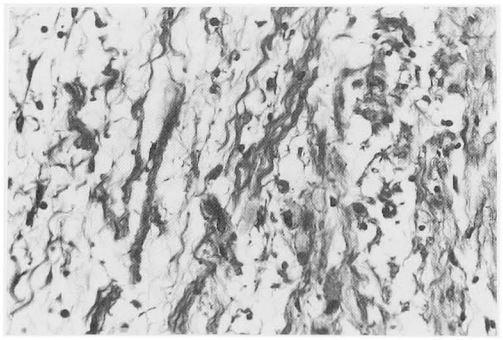

Fig. 3. Histologic appearance of the tumor shows fusiform tumor cells in a matrix (H & E stain).

(Fig. 4, A and B). Thus, the tumor was diagnosed as a solitary neurofibroma. The patient remains well without any signs of tumor recurrence.

DISCUSSION

A neurofibroma and a schwannoma

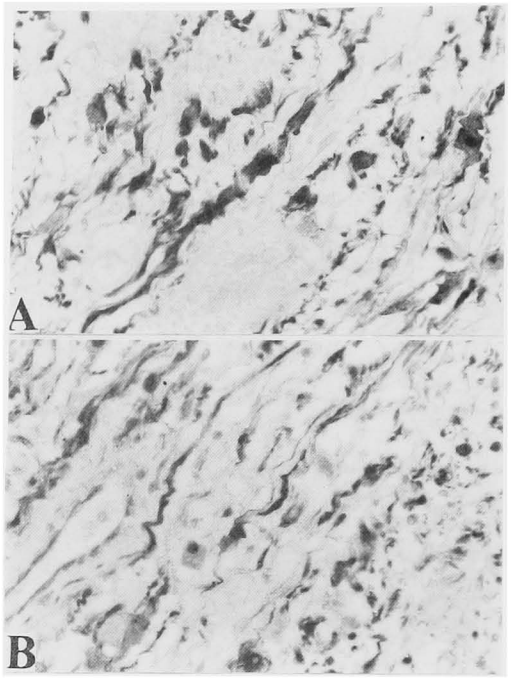

Fig. 4. Immunohistochemical demontsration of S-100 (A) and NSE (B). The tumor cells show an intense positive immunoreaction in the cytoplasm (PAP immunoperoxidase stain without counter stain).

(neurilemoma) are benign nerve sheath tumors, originating from the Schwann cells. Although there is confusion relating to differentiation between a neurofibroma and a schwannoma, a neurofibroma is distinct from a schwannoma in the following points. (i) The former lacks the compact arrangement of cells in the Antoni type A tissue and delicate single cell thick fascicles of the Antoni type B tissue of a schwannoma[7] (ii) Alcian blue stains for acid mucopolysaccharides are negative in a schwannoma but positive in a neurofibroma[7]. (iii) A neurofibroma contains fewer S-100 protein positive cells than a schwannoma[8].

S-100, a dimetric calcium binding protein, composed of different combination of α and β subunits, mainly locates in glial and Schwann cells[9]. NSE, a glycolytic dimetric enzyme, locates in neurons and neuroendocrine cells[10]. Immunohistochemical demonstration of S-100 and NSE is valuable in the diagnosis of

a nerve sheath tumor. Unbalanced expression of S-100 subunits is observed in malignant nerve sheath tumors. Hayashi et al. observed an increased expression of the S-100 α subunit in the malgnant part of neurofibromas[11]. They concluded that the S-100 subunit immunoreactivity is a good marker for malignant schwannomas.

As described above, we could not distinguish between a neurofibroma and a schwannoma in the previous literature. Das Gupta et al. considered solitary neurofibromas and schwannomas (neurilemomas) as the same entity, and they reviewed 303 such cases[12]. In that report, most tumors occurred in the head and neck, and extremities. On the other hand, only two of these cases (0.66 %) were retroperitoneal origin. Surgical excision of the tumor is the choice of treatment[6]. Careful search for the coexistence of a malignant tumor is necessary, because 16 % of the patients with a solitary neurofibroma were reported to have an associated malignant tumor[12].

It remains to be elucidated whether the clinical course of a solitary neurofibroma differs from that of a schwannoma.

REFERENCES

1) Gonzalez-Angulo A and Reyers HA: Neurofibromatosis involving the lower urinary tract. J Urol **89** :804-811, 1963
2) Winfield HN and Catalona WJ: An isolated plexiform neurofibroma of the bladder. J Urol **134**: 542543, 1985
3) Freund ME, Crocker DW and Harrisier JH Neurofibroma arising in a solitary kidney. J Urol **98**: 318-321, 1967
4) Ravich A : Neurofibroma of the ureter : report of a case with operative and recovery. Arch Surg **30**: 442-448, 1935
5) Elliott FG, Eid TC and Lakey WH: Genitourinary neurofibromatosis : clinical significance. J Urol **125**: 725-727, 1981
6) Carpenter WB and Kernohan JW: Retroperitoneal ganglioncuromas and neurofibromas: a clinicicopathological study. Cancer **16**: 788-797, 1963
7) Harkin JC and Reed RJ: Solitary neurofibroma (Solitary nerve sheath tumor). In : Tumors of the peripheral nervous system. Edited by Hasrkin JC and Reed RJ. Armed Forces Institute of Pathology, Fasc. 3, pp. 51-59, Washington, D.C., 1968
8) Weiss SW, Langloss JM and Enzinger FM: S-100 protein in the diagnosis of soft tissue tumors with particular reference to benign and malignant Schwann cell tumors. Lab Invest **49**: 299-308, 1973
9) Stefansson K, Wollmann RL and Moore BW Distribution of S-100 protein outside the central nervous system. Brain Res **234**: 309-317, 1982
10) Kato K, Suzuki F and Semba R . Determination of brain enolase isozymes with an enzyme immunoassay at the level of single neurons. J Neurochem **37**: 998-1005, 1981
11) Hayashi K, Tak ahashi K, Sonobe H, Ohtsuki Y and Taguchi K:The distribution of alpha and beta subunits of S-100 protein in malignant Schwannomas arising from neurofibromatosis of von Recklinghausen's disease. Virchows Arch A **411** : 515-521, 1987
12) Das Gupta TK, Brasfield RD, Stbong EW and Hajdu SI: Benign solitary Schwannomas (Neurolenomas). Cancer **24**: 355-366, 1969
13) Steers WD, Hodge GB, Johnson DE, Chaitin BA and Charasangavej C: Benign retroperitoneal neurilemoma without von Recklinghausen's disease : a rare occurrence. J Urol **133**: 846-848, 1985

(Accepted for publication July 25, 1988)

和文抄録

孤立性後腹膜神経線維腫の1例

神戸大学医学部泌尿器科学教室（主任：守殿貞夫教授）
石川二朗，守殿貞夫
神戸大学医学部第2病理学教室（主任：杉山武敏教授）
前田盛，杉山武敏
原泌尿器科病院（院長：原信二）
原勲，武市佳純，大前博志，原信二

後腹膜に原発し，von Recklinghausen 氏病の徴候のない孤立性神経線維腫の1例を報告する．診断上，もう一つの神経鞘由来の神経鞘腫との鑑別が問題で，腫瘍の組織化学染色および免疫組織化学染色が孤立性神経線維腫の診断に有用であった．

（泌尿紀要 **35**：1157-1160，1989）