[症例報告]Schwannomas in Ryukyu University Hospital 1983-1991

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ABSTRACT

From 1983 to 1991, total 3 cases of schwannomas have been registered in the Department of Dermatology of University of the Ryukyus Hospital. They comprised only 0.026% of total out-patients and 0.11% of new out-patients in a year. All the cases were ordinary types of schwannoma. The tumor occurred on the right thigh of a 52-year-old woman measured 20 × 15 × 12cm and weighed 2250g; it is probably the largest schwannoma reported on the extremities in Japan.

INTRODUCTION

Schwannomas are known to be one of the common soft tissue tumors. According to a survey on their incidence, they are the third most common following angiomas and lipomas, and comprise roughly 10% of the benign soft tissue tumors as a whole[1]. They occur practically in every anatomical region. Among the schwannomas of dermatological interest, sizes range from 0.2cm to 20cm in maximum diameter. Approximately 90% of the reported cases so far have the maximum diameters less than or equal to 5cm. Only 12 cases whose maximum diameters are bigger than 5cm, have been reported in Japan since 1973. In our out-patient clinic, total 3 cases of schwannomas have been seen since 1983. In this study, the brief outline of each case is portrayed in a chronological order of registration, and a clinical observation is discussed.

CLINICAL MATERIAL

During the period from 1983 to 1991, total 3 cases of schwannomas were seen at the De-

Table 1. Summary of three cases of schwannomas

Case No.	Age (years)	Sex	Date of lst.visit	Site	Size (cm)	Duration prior to lst.visit (years)	Histological diagnosis
1	43	M	1988	left leg	2×2×2	3	Antoni A+B ordinary type
2	72	F	1988	back	0.7×0.7×0.3	3	Antoni B ordimnary type
3	52	F	1990	right thigh	20×15×12	10	Antoni A+B ordinary type

-partment of Dermatology of the University of the Ryukyus Hospital, 2 cases in 1988 and 1 case in 1990. They comprised only 0.026% of total out-patients and 0.11% of new out-patients in a year. Table 1 shows their brief outlines.

Case 1 : A 43-year-old man was first seen, in July 1988, with a lump on the posterior aspect of his left leg. He had noticed it 3 years earlier for the first time. Since then it gradually increased in size, though remaining asymptomatic throughout its growth. Physical examination revealed a moderately firm, subcutaneous movable mass measuring 2 × 2cm. The tumor was easily enucleated. Histological findings were as follws: the tumor was encapsulated with fibrous membranes. Inside the mass, proliferation of the spindle-shaped cells was observed, whose nuclei were elongated. Some of those cells were arranged in so-called pallisading arrangement. In some areas, aggregations of clear round cells with mucinous degeneraton were seen. The histological diagnosis was an ordinary type of schwannoma conposed of two types of tissue, Antoni types A and B. The patient has been well and without recurrence for about 4.5 years following the excision.

Case 2 : A 72-year-old woman was first seen, in November 1988, with a bean-sized nodulus on the back. She noticed it 3 years earlier for the first time. She told us that it was asymptomatic and the size was relatively stable. On physical examination, the tumor was skin-colored, soft and half dome-like with a smooth surface. It measured 7×7×3mm. Histological findings were as follows : under the epithelium, loose, edematous, hypervascular, and relatively hypocellular tissues were seen. The constituent cells were long, spindle-shaped with nuclei of variable shapes. No mitoses were detected anywhere. The histologic diagnosis was Antoni B type tissue of an ordinary type of schwannoma. The patient has been well for about 4 years after the operation.

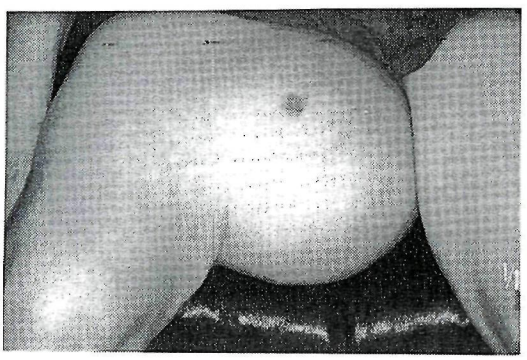

Fig. 1. Clinical appearance of Case 3 in April 1990, showing the anterior aspect of a big subcutaneous tumor on the medial femoral region of the right thigh measuring 20×15×15cm.

Case 3 : A 52-year-old woman was first seen, in April 1990, with a man's head sized lump on her right thigh. She noticed it about 10 years earlier for the first time. It was henegg sized at that time. Since then it had gradually increased in size to that of a man's head.

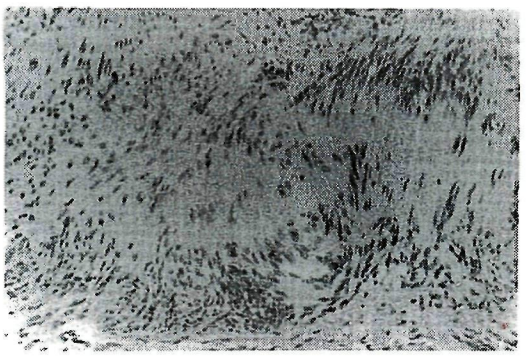

Fig. 2. Histopathology of Case 3. The section taken from the central area shows hypercellular portions composed of spindle-shaped cells forming so-called Verocay bodies, i.e., Antoni type A tissue. Other specimens taken from the peripheral area show hypocellular portions of edematous stroma containing few haphazardly arranged cells with nuclei of variable shapes, i.e., Antoni type B tissue. (Hematoxylin-eosin stain, ×200)

It remained asymptomatic throughout its growth. Physical examination revealed no abnormalities except a big subcutaneous tumor on the medial femoral region of the right thigh, which measured 20 × 15 × 15cm (Fig. 1). It was roughly egg-shaped, normal skin colored, elastic hard with a smooth surface, well-demarcated, slightly movable, and not adhesive to the surrounding tissues. Neither sensory abnormalities nor lymph node enlargement were detected on her body.

On admission, laboratory findings including blood analysis, chemisry tests, serologic tests and urinalysis were essentially within normal limits. However, a CT examination as well as an ultrasonograph of the femoral region revealed a well-demarcated, almost homogeneous, solid mass with small, round shadows of increased density that suggested some cystic degeneration inside the mass. The boundary between the mass and the femoral muscles was clear. No invasion to the muscles or to the surrounding tissues was detected.

Based upon the above findings, we provisionally diagnosed the tumor as a benign soft tissue tumor, and completely excised it. The removed tumor was encapsulated within semitranslucent membranes. It measured 20×15×12 cm and weighed 2250g. On sectioning the tumor perpendicular to its long axis, the cross sections revealed yellow to pearl gray, multilobulated, intracapsular tissues with partially cystic degeneration and hemorrhages.

Histopathological findings were as follows: in general, based upon observation of all the specimens taken from various areas of the tumor, the specimens from the central areas were significantly hypercellular; whereas those from the peripheral areas included hypocellular portions with various degeneration. The hypercellular portions were composed of spindle-shaped cells whose nuclei were elongated and tightly packed. They were arranged in many areas in a streaming fashion, and formed so-called Verocay bodies in some areas (Fig. 2). Essentially no mitotic figures were detected in the specimens from these areas. From these findings, it was concluded that the histological diagnosis was Antoni type A schwannoma. On the other hand, the hypocellular portions were composed of edematous stromas that contained relatively few, haphazardly arranged cells with nuclei of variable shapes. In places, fatty degeneration, cystic degeneration, hemorrhages and necrosis were observed. Again, there was no mitosis. These findings suggested that the histological diagnosis for these areas was Antoni B type schwannoma. In order to confirm the diagnosis, other stains such as immunoperoxidase stains against S-100 protein and neuron-specific enolase were utilized[2]. No contradictory results were obtained. We therefore concluded that the final diagnosis for this tumor was a mixture of two types of tissue Antoni types A and B of an ordinary type of schwannoma.

The postoperative course of the patient was

uneventful; her general status as well as the local wound healing were good. She has been well for approximately 3 years after the operation without a recurrence.

DISCUSSION

It is widely accepted today that schwannomas are tumors of nerve sheath origin. They arise most commonly from either cranial or peripheral nerves, and develop practically in every anatomical region. According to the statistic in 1974 about benign soft tissue tumors in Japan by Enjoji et al., schwannomas are the third most common, following hemangiomas and lipomas and comprise about 10% of benign soft tissue tumors as a whole[1]. However, as for the incidence, there is enough room for controversy: some authors disagree with the figure and suggest that the actual incidence should be higher, indicating a statistical inadequacy of the study[3]. In our office, there is not enough accumulation of schwannoma cases to make any comment on this issue.

In our small group of the 3 cases, the anatomical distribution of the tumors was as follows: one was seen on the trunk and two were seen on the lower extremities. In 1969, Das Gupta et al. studied 303 patients with benign solitary neurogenic tumors in the U.S. and reported the anatomical distribution of schwannomas as follows : 44.8% of them were located in the head and neck region, 19.1% on the upper exremity, 8.5% on the trunk, 13.5% on the lower extremity and 13.8% in various unusual sites[4]. On the other hand, in Japan, Enjoji et al. reported, in the above mentioned paper, the following distribution of 825 casea of schwannoma: 29.3% of them were located in the head and neck region, 20.5% on the upper extremity, 22.5% on the trunk, 21.8% on the lower extremity and 5.3% in miscellaneous regions.

These statistics differ. However, it is not clear what kind of factors (e.g. races, statistical biases, etc.) are involved. There has been no study which compares the incidence of such tumors among the races.

In 1989, Iwashita reported an almost identical distribution of schwannomas as that of Enjoji et al. on the basis of a review of 1271 cases of schwannoma registered to the Histopathology Office of Kyushu University from 1951 to 1987 : 26.6% were distributed in the head and neck region, 24.0% on the trunk, 24.5% on the upper extremity and 24.2% on the lower extremity[5]. In summarizing these statistical figures, it seems safe to say that if the surface of the whole body is divided into these 4 major regions, the frequencies in the respective regions are almost the same, namely 20-25%.

Male-female ratio of schwannomas in our small group was one to two favoring females. However, Iwashita reported that sex of the patients was not an important factor in schwannomas: it occurs equally on both sexes with the ratio of approximately one to one[5]. Many authors have given unanimous approvals to this statement[1,3,6].

Ages at onset of schwannomas were reported to range from 2 to 89 years with a mean of 46[5]. Das Gupta's report as well as others almost coincide with it[1,4]. Our small group showed a mean age of 55.7, which is slightly higher.

As briefly mentioned in the introduction, maximum diameters of reported cases of schwannoma range from 0.2cm to 20cm with a mean of 3.1cm[5]. Soul stated in 1954 that those that arose on the extremities never exceeded 6cm in diameter[7]. Harkin et al. stated in 1968 that they would rarely become bigger than 8cm[8]. From these assertions, it seemed reasonable to us to agree with defining giant schwannomas as those whose diameters were bigger than 5cm[9]. Using this working definition, we studied the literature of giant schwannomas on the extremi-

-ties in Japan from 1973 to 1992. Approximately 90% of the reported cases so far had the maximum diameters less tham or equal to 5cm[1,3,4,5]. Of those whose maximum diameters are bigger than 5cm, only 12 cases in total have been reported since 1973 in Japan[9]. The biggest one, 20×12×10cm, that occurred on the lower extremity was reported by Ichise et al. in 1973[10]. One of the cases in this paper described as case 3 was slightly bigger, i.e., 20×15×12cm; this is probably the biggest schwannoma on the extremities in Japan. Our small group showed an average size of 7.6cm. It is significantly bigger than the average size mentioned above (3.1cm). This is clearly due to the large contribution of case 3.

Most of schwannomas arising on the extremities are small. One reason for this may be that they are easily recognized from outside either by the patients or by their family members. On the other hand, for those that arise somewhere in the deep anatomical regions of the body such as the pelvic cavity or the retroperitoneum, it is usually not until they become big enough to cause some symptoms that the patients initially become aware of them. Incidentally, the biggest schwannoma reported in 1978 in Japan arose from the retroperitoneum, which measured 30×30cm and weighed about 5kg to our knowledge[11]. The true reason why case 3 showed such big dimensions is still unknown. However, we speculated that some peculiar character of the patient must have played a role in her prolonged first visit to a hospital. She had never shown the tumor even to her husband before the first visit and refused to let him know her hospitalization even after her discharge, probably with some unknown anxieties.

Iwashita mentioned in the above paper that the relationships between the tumors and the nerves involved in them or responsible for the formation of the tumors were clearly discussed only in 22% of the registered cases[5]. Manabe et al. reported in 1987 that the nerves involved in the tumors were identified during operation in 70% of 105 schwannoma cases[3]. They speculated, however, that in the remaining 30% of the cases with unidentified nerves, minute muscular or cutaneous branches must be responsible. We were not able to detect the nerves involved in case 3 during the operation and reached the same speculation.

There have been several proposals regarding subtyping of schwannomas. According to Iwashita's attempt, they are classified into 7 subtypes: ordinary, cellular, plexiform, pigmented, myxomatous and organoid types[5]. The respective frequencies of these subtypes are 73% for ordinary type, 19% for degenerated type, 3% for plexiform type, 2% for organoid type, 1% for cellular type, and 1% for myxomatous type. In our small group, all were ordinary types.

To compare the frequencies of schwannomas in out-patient clinics of dermatology of university hospitals in Japan, we looked for such statistics in the literature. Very few reports have been seen regarding this issue. One such report was presented indirectly by the Department of Dermatology, Kurume University, School of Medicine: 5 cases of schwannomas were seen from 1975 to 1985[12]. The figure is slightly bigger than ours. Other institutes including surgical departments reported much bigger figures. One reason for this may be that patients with schwannomas of small sizes tend to visit a surgical clinic rather than a dermatology clinic of a university hospital, eventually resulting in a higher incidence in the former. However, it is not clear whether schwannomas are rare in Okinawa. Further studies are needed such as a nation-wide survey on the incidence.

REFERENCES

1) Enjoji, M., Iwasaki, H., and Komatsu, K.: Benign soft tissue tumors in Japan-A statistic analysis of 8086 tumors, Gan no Rinsho, 20 : 594-609, 1974 (in Japanese).

2) Weiss, S. W., Langloss, J. M., and Enzinger, F. M.: Value of S-100 protein in the diagnosis of soft tissue tumors with particular reference to benign and malignant Schwann cell tumors, Laboratory Investigation, 49 : 299-308, 1983.

3) Manabe, M., Kawaguchi, T., Amino, K., Matsumoto, S., Kitagawa, T., Machinami, M., Totake, M., Huruya, K., Wada, N., Isobe, Y., and Tako, M.: A clinico-pathological study on 105 cases of schwannoma developed in peripheral nerves, Rinsho Seikeigeka, 22 : 1035-46, 1987 (in Japanese).

4) Das, Gupta, T. K., Basfield, R. H., Strong, E. W., and Hajdu, S. I.: Benign solitary schwannomas (neurilemomas), Caner, 24 : 355-66, 1969.

5) Iwashita, T.: Neurilemomas of the soft tissues-An analysis of 1271 tumors in an attempt at subtyping, Fukuoka Ishi, 80 : 355-67, 1989 (in Japanese).

6) Hu, F.: Neurilemoma, in Demis DJ, Thiers BH, Smith EB, Burgdorf WHC (eds) : Clinic-al Dermatology, Lippincott, Philadelphia, 1991, unit 25-2.

7) Soul, E. H.: Primary soft tissue tumors of the extremities; Classification, histogenesis and incidence, Acad. Ortho. Surg., 11 : 3-11, 1954.

8) Harkin, J. C., and Reed, R. J.: Tumors of the peripheral nervous system, in Atlas of Tumor Pathology, 2nd Series, Fascicle 3, p29-51, Armed Forces Institute of Pathology, Washinton, D. C., 1968.

9) Nakajima, K., Koshino, T., Morii, T., Ejima, M., and Kushida, K.: Giant schwannoma arising from the tibial nerve-Two case reports, Kantou Seisaishi, 18 : 365-69, 1987 (in Japanese).

10) Ichise, T.: Two cases of neurilemomas arising from the peripheral nerves, Kantou Seisaishi, 4 : 56-60, 1968 (in Japanese).

11) Hiramatsu, Y., Shidahara, M., Yonezawa, Y., Shimizu, K., and Sekiba, K.: A case of retroperitoneal neurilemoma and a statistical observation of 48 cases in Japan, Sanka to Fujinka, 49 : 110-6, 1982 (in Japanese).

12) Tanaka, H., Hachisuka, H., Mori, O., and Sasai, Y.: Re-investigation about tumors of neural tissues with light microscopy, Japanese Journal of Clinical Dermatology, 39(1) : 61-4, 1985 (in Japanese).