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Case Report
Extrarenal Angiomyolipoma : Report of a Mediastinal Case and Review of the Literature

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ABSTRACT. A mediastinal angiomyolipoma was excised from a 17-year-old man. It was composed of capillaries and cavernous hemangioma-like vessels, smooth muscle cells and mature fat cells. The histology was somewhat different from that of ordinary renal angiomyolipoma. Extrarenal angiomyolipoma has been also reported under other terms. It seems extremely rare. To our knowledge, only 22 cases of such examples have been reported in English literature. Here, we present our case with histological comparisons between renal and extrarenal varieties. A possible histogenesis is also discussed.

Key words : Extrarenal Angiomyolipoma — Mediastinum

Angiomyolipomas are defined as tumors or tumor-like lesions composed of blood vessels, smooth muscle cells, and fat cells. The majority of them originate in the kidney, and approximately half of these lesions are associated with tuberous sclerosis. Extrarenal angiomyolipomas appear to be extremely rare, and are mostly reported under the various designations such as benign mixed mesodermal tumor[1], hamartoma[2], benign mesenchymoma[3], or cavernous hemangioma[4]. Benign mixed tumors of adipose tissue and other mesenchymal elements may sometimes be referred to as a more ambiguous term of benign mesenchymoma, but more precise descriptive names such as angiomyolipoma or lipomyohemangioma are apparently preferred.

Recently, we have encountered a case of mediastinal angiomyolipoma in which the cavernous hemangioma-like vascular element was predominant over the tortuous arteriolar element. Here, we describe our case with the literature review, and try to compare the histology of renal and extrarenal angiomyolipomas. A possible histogenesis is also discussed.

CASE REPORT

A 17-year-old man with an abnormal mediastinal shadow found on chest

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x-ray at the ages of two and fourteen years was admitted to the Kawasaki Medical School Hospital. He had been asymptomatic. There was no abnormalities suggestive of tuberous sclerosis such as mental retardation, epilepsy, and sebaceous adenoma. A chest x-ray and computed tomographic (CT) scan of the mediastinum revealed a mass to be localized in the right upper-medial part of the mediastinum. The comparison of the chest x-ray at 14 and 17 years of age did not show any change in size. The mass was of high density but non-homogeneous on CT scan. Angiography revealed the superior vena cava (SVC) and left brachiocephalic vein (BCV) to be dilated with the former being compressed from the anterior side. No irregularity of the venous wall was noted. No other abnormalities were found with routine physical and laboratory examinations. Operation was done on the eleventh hospital day. The tumor was mainly located in the superior mediastinum anterior to the SVC and BCV but in part encased them as well as right phrenic nerve. It was also firmly adherent to the upper portion of the pericardium. Therefore, the complete separation from SVC and BCV was not possible. The tumor was excised with an involved part of pericardium and adjacent thymus, but the portion near SVC and BCV was left untouched.

PATHOLOGICAL FINDINGS

The excised tumor measured $6 \times 5 \times 2$ cm and was reddish brown and resilient in consistency. Cut surfaces showed variegated appearances. Numerous vessel-like spaces, hemorrhagic areas and gray fibrous and yellowish fatty tissue were intermingled. The thymus abutted on the tumor with clear border.

Microscopically, it was composed of blood vessels, smooth muscle cells, fat cells and supporting stroma of fibrous connective tissue (Fig. 1). These components were intimately admixed in various proportions. The blood vessels displayed a great variety in size, structure and arrangement. Many of them were venous in appearance and mostly dilated. Thin walls were composed of fibrous tissue as well as a small amount of smooth muscles but no elastic fibers were discernible. A relatively small number of normal-looking capillaries, venules and small arteries were also scattered in the tumor. In addition to these vessels, large anastomosing channels, the walls of which were irregular in thickness and mainly composed of smooth muscle cells, were seen (Fig. 2). Their walls lacked elastic tissues as well. Endothelial cells were flattened and showed no mitotic figures in all blood vessels. Smooth muscle cells in the stroma around them were present singly or as small groups or bundles. The smooth muscles of vascular walls merged into the extravascular muscular structures. Cellular atypism or mitosis was not seen. Fat cells had a large clear cytoplasm and an eccentric nucleus. All adipocytes were mature and no mitoses were seen. Small-sized lymphocytes, plasma cells, lymph follicles, and hemosiderin-laden macrophages were scattered in the stroma.

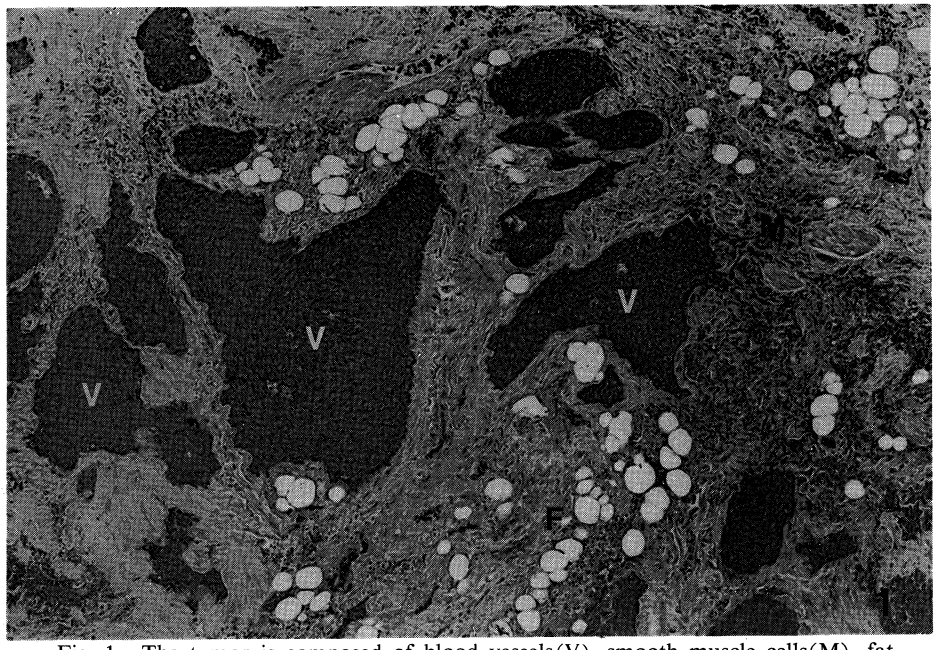

Fig. 1. The tumor is composed of blood vessels(V), smooth muscle cells(M), fat cells(F), and stromal fibrous tissue. Many of the the vessels show cavernous hemangioma-like appearance. (H-E, ×60)

DISCUSSION

Benign mesenchymal tumors are common in surgical pathology. Many of them are composed solely of fibrous tissue, adipose tissue, smooth muscle, striated muscle, lymph vessel, blood vessel, bone, cartilage, or myxomatous tissue. In some, however, more than two elements may exist admixed in the same tumors. Only those tumors showing the differentiation into two or more unrelated forms of mesodermal tissue other than fibrous tissue, are called mesenchymomas[5,6]. Their tissues are usually mature but abnormal in arrangement. Fat cells, blood vessels, and smooth muscle cells are the most common elements in these mesenchymomas[3]. Those tumors may be named more descriptively. For instance, tumors having adipose tissue, blood vessels, and smooth muscles together can be designated as "angiomyolipomas". Depending upon the proportion of each element, other names may be employed; namely lipomyohemangioma or myoangiolipoma.

Renal angiomyolipomas are by far the commonnest but extrarenal cases are also known. Keeping the variety in terminology in mind, we were able to collect 22 examples from English literature[1-4,7-9]. The uterus, extremities, liver, mediastinum, perirenal tissue, hard palate, and eyelid were the anatomical sites. Pertinent clinical and pathological data are summarized in Table 1. The

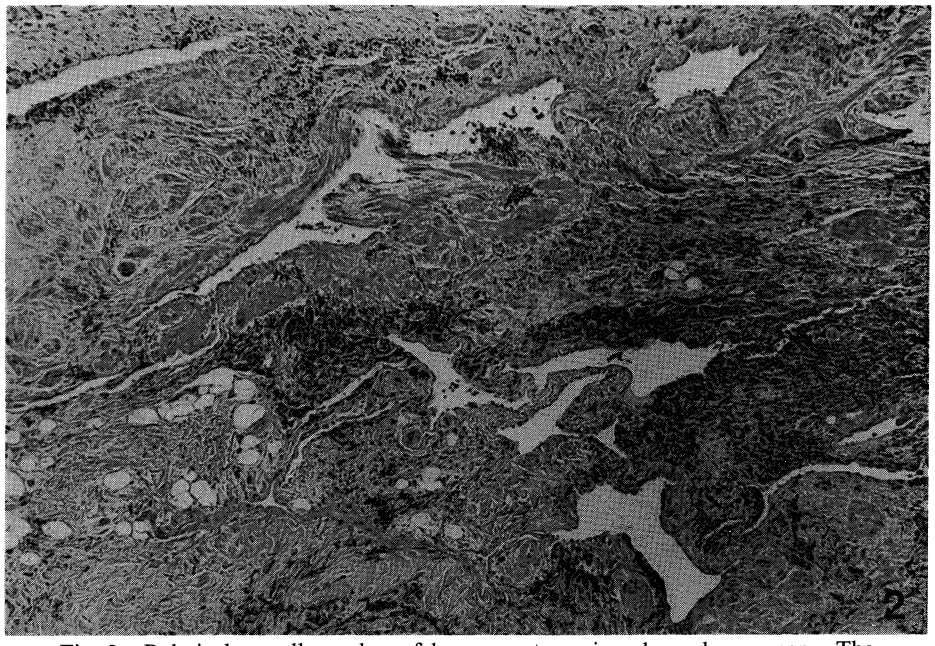

Fig. 2. Relatively small number of large anastomosing channels are seen. The smooth muscles of vascular walls merge into the extravascular muscular structures. (H-E, ×60)

histology of renal angiomyolipoma has been well-documented and is characterized by three elements; adipose tissue cells, thick-walled tortuous blood vessels and sheets of smooth muscles. Each component varies in the amount and histological appearance. Although the blood vessel component also displays great variation in type, size, arrangement and structure, the major portion consists of large arterial type vessels which are extremely tortuous, reminiscent of cirsoid aneurysms. The media are frequently of variable thickness and structure. They sometimes consist of smooth muscle, at other times of collagen and muscle, and still others of hyalinized collagenous connective tissue. Most vessels have little or no elastic tissue, but consist of closely packed immature appearing smooth muscle cells arrayed circumferentially.

In view of the presence of tortuous arterial type vessels, angiomyolipoma was thought unique to the kidney. In fact, Allen[10] stated that histologically identical tumors probably do not occur in any other location. In our literature review, however, they do exist, and were found in the liver, uterus, and hard palate[1,2,7–10]. In other extrarenal cases, the histology of the vascular elements was not well-described. For unknown reasons, however, cavernous hemangioma-like vascular element seems to be exaggerated in mediastinal cases, as is in our case, and it was called cavernous hemangioma of the mediastinum. It must

Table 1 : Extrarenal Angiomyolipoma

Case	Sex	Age at onset	Age at treat. or autopsy	Tumor size	Anat. site	Type of treat.	Recur.	TS	Vascular element	Term applied
1. Present case	M	2yr.	17yr.	6×5×2 cm.	Mediast.	Exc.	(−) 17mo. later	(−)	Cavernous	Angiomyolipoma
2. Gindhart T.D.[4]	M	?	38yr.	4cm.	Mediast.	Exc.	(−) 8mo. later	(−)	Cavernous	Cavernous hemangioma
3. Le Ber M.S.[3]	F	?	7yr.	8cm.	Mediast.	Exc.	?	(−)	NCD	Benign Mesenchymoma
4. Le Ber M.S.[3]	M	?	11yr.	Lge.	Above kidney	Exc. Nephrect.	?	(+)	NCD	Benign mesenchymoma
5. Le Ber M.S.[3]	F	?	13mo.	?	Elbow region	Exc.	(+)	(−)	NCD	Benign mesenchymoma
6. Le Ber M.S.[3]	F	Birth	14yr.	13×8×4.5 cm.	Thigh & Knee	Radiat. Exc.	(+)	(−)	NCD	Benign mesenchymoma
7. Le Ber M.S.[3]	F	Birth	14yr.	?	Ankle	Exc.	?	(−)	NCD	Benign mesenchymoma
8. Le Ber M.S.[3]	F	Birth	14mo.	?	Eyelid	Attempt. Exc.	Died at op.	(−)	NCD	Benign mesenchymoma
9. Le Ber M.S.[3]	M	?	4yr.	Extens.	Leg	Exc.	?	(−)	NCD	Benign mesenchymoma
10. Le Ber M.S.[3]	F	?	14yr.	7×4×2 cm.	Forearm	Exc.	(−)	(−)	NCD	Benign mesenchymoma
11. Le Ber M.S.[3]	F	?	12yr.	?	Retroper.	Exc.	?	(−)	NCD	Benign mesenchymoma
12. Demopoulos R.I.[1]	F	Few mo. ago	60yr.	12cm.	Uterus	Hysterec. ?	?	(−)	*	Benign mixed mesodermal tumor
13. Demopoulos R.I.[1]	F	Several mo. ago	48yr.	30cm.	Uterus	Hysterec. ?	?	(−)	*	Benign mixed mesodermal tumor
14. Demopoulos R.I.[1]	F	6mo. ago	49yr.	14cm.	Uterus	Hysterec. ?	?	(−)	*	Benign mixed mesodermal tumor
15. Demopoulos R.I.[1]	F	Several mo. ago	39yr.	4cm.	Uterus	Hysterec. ?	?	(−)	*	Benign mixed mesodermal tumor
16. Demopoulos R.I.[1]	F	Several mo. ago	44yr.	16cm.	Uterus	Hysterec. ?	?	(−)	*	Benign mixed mesodermal tumor
17. Demopoulos R.I.[1]	F	Several mo. ago	59yr.	6cm.	Uterus	Hysterec. ?	?	(−)	*	Benign mixed mesodermal tumor
18. Demopoulos R.I.[1]	F	Several yr. ago	61yr.	2cm.	Uterus	Hysterec. ?	?	(−)	*	Benign mixed mesodermal tumor
19. McKeithen W.S.[2]	F	?	62yr.	2×1.3×1.3 cm.	Uterus	Hysterec.	?	(−)	*	Hamartoma
20. Ishak K.G.[7]	F	?	66yr.	2cm.	Liver	——	——	(−)	*	Angiomyolipoma IAA
21. Ishak K.G.[7]	M	?	57yr.	1cm.	Liver	——	——	(−)	*	Angiomyolipoma IAA
22. Pounder D.J.[8]	M	?	65yr.	3cm.	Liver	——	——	(−)	*	Angiomyolipoma IAA
23. Gutmann J.[9]	M	At least 2yr. ago	39yr.	1×0.6×0.6 cm.	Hard palate	Exc.	?	(−)	*[10]	Angiomyolipoma

Treat. : Treatment Anat. : Anatomical Recur. : Recurrence TS : Tuberous Sclerosis NCD : Not Clearly Described
Exc. : Excision IAA : Incidental At Autopsy * : Indistinguishable From Renal Angiomyolipoma

be remembered that these are also angiomyolipoma in morphological sense.

The nature of this tumor is still unclear, although many authors consider it a tissue malformation or hamartomatous growth rather than a true neoplasm. One may speculate the existence of pluripotential mesenchymal cells, which may become neoplastic to differentiate into endothelial cell, smooth muscle cell, lipocyte, and/or fibroblast. It is, however, quite hard to believe that such neoplastic pluripotential mesenchymal cells differentiate enough to form well-organized arterial or venous structures. In addition, supportive evidence for hamartomatous nature may be provided by the fact that although benign mesenchymomas may recur if excised incompletely[11], malignant transformation with fully-developed blood vessels is not known.

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