症 例 報 告

SYMPATHICOGONIOMA OF THE ADRENAL MEDULLA: REPORT OF A CASE

by

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A primary malignant tumor of the adrenal medulla is rarely encountered. About 300 cases of it have been reported in foreign countries since PARKER and MERCHANDT (1880) and a little over 40 cases in Japan since FUJIYOSHI (1909). It is usually divided into two types according to the preponderant cells: sympathicogonioma and sympathicoblastoma. The latter is composed of sympathoblasts, which present a higher degree of differentiation than sympathogonia, and the tumor is practically less malignant than sympathicogonioma, and most commonly occurs along paravertebral symbathetic ganglia, but rarely in adrenal.

Sympathicogonioma is the most common tumor found in the sympathetic nervous system, composed of the primitive cells, and occurs almost always in the adrenal. It is almost entirely found in the early childhood; about 80 per cent of the cases occuring in children, less than 6 years of age, and 30 per cent in infants within 1 year after nativity. Of 40 cases, reported by Lewis and Geschickter, only 5 patients were more than 15 years old, including one in a man of 45 years of age. However, as pointed out by Karsner, it is probable that the tumor of these cases should in present knowledge be classified as sympathicoblastoma. Practically it frequently metastatizes to the liver or to the skeletal system, especially the skull and long bones, and invades adjacent structures in the early stage.

In our present case of sympathicogonioma, the patient presented the signs and symptoms of intestinal obstruction by the compression of a large tumor in the left hypochondrium, but metastases were not evident.

CASE REPORT

K. M., 9 months old female infant, was admitted to Pediatric Clinic of our Hospital on Jan. 20, 1954, complaining of obstinate constipation and vomiting.

She had grown in excellent health till about 3 months before admission, then she gradually became constipated. At the end of 1953, only a little amount of stool was discharged every 5 to 7 days by repeated clysters. Distension of the

abdomen and remittent fever appeared additionally, and she began to vomit after every meal. At last, as the vomiting increased in frequency to ten or more times in a day and she became remarkably emaciated, she was admitted to Pediatric Clinic of our Hospital. Since then her constipation improved a little and defecation appeared every 3 or 4 days, but in a small amount.

Vomiting temporarily disappeared. At that time, a large tumor was recognized in the left hypochondrium. On roentgenologic examination, giving contrast medium by oral and also by anal route, a passage failure caused by the large tumor was proved in the region of flexura lienalis. On the pyelogram, by means of intravenous injection of Sugiuron (33% solution of di-jod-methyl-selidum acid potassium) the shadows of the renal pelvis and ureter appeared normally on the right side, but failed completely on the left side. Thus, she was referred to our clinic for surgical treatment on Jan. 26, 1954.

CLINICAL FINDINGS : On admisstion, the patient was very weak and remarkably emaciated. The skin was pale and dry. The tongue was coated and dry. No enlargements of lymph nodules. Neither swellings nor deformities of the skull and extremities were externaly noticeable. Exophthalmos, ecchymosis or edema of eyelids were not proved. A moderate anemia was revealed on blood examination. But leucocytosis and hyperglycemia were not evident.

Urinary proteins was positive and a few leucocytes were found in urine sediment. But hematuria and reduction in daily urine amount had never been remarked. Occult blood test of feces indicated positive.

The abdomen was distended diffusely and strikingly, but the abdominal wall was soft on palpation. Neither signs of peritonitis, nor violent peristalsis were noticeable. In the left hypochondrium, we could find a tumor which was larger than a fist, nodular and firm elastic. The upper, medial and lower borders of the tumor were relatively sharp, but the lateral border was hidden by the left costal margin. The tumor extended upward to about 1.5 cm below processus xyphoideus, medially to about 2 cm right beyond the midline and downward to about 1.5 cm below umbilicus, and was not movable with respiration and also passively. A part of the tumor could be grasped bimanually and such an indentation as seen in splenomegalia was not palpable at any border. On percussion of the abdomen, the area covering the tumor was dull.

Ascites was not recognizable. The sharp and soft edge of the liver was palpable at about 1 cm under the right costal margin and the surface of the liver was smooth.

Not only clinical findings, but also the roentgenological findings, especially the pyelographic pictures in this case seemed to be satisfactory to make a diagnosis of renal tumor, although hematuria and decrease in urine amount had never been remarked.

From the next day of admission to our clinic, the patient began to vomit again and was attacked with fever. She finally vomited a feces-like fluid.

OPERATION : Under local anesthesia a pararectal section was made and the peritoneum was opened. There was found no ascites in the abdominal cavity.

The liver and mesenterial lymph nodules seemed to be normal in appearence.

The tumor was as large as a child's head in size, and situated retroperitoneally at the lateral portion of the descending colon. It was reddish brown nodular, soft elastic or partially cystic, rich in blood vessels on the surface and encapsulated by a fibrous thin menbrane. It arose from the upper portion of the left kidney and covered the kidney entirely, which was shifted by compression back- and downward. The right border of the tumor reached the right kidney over the backbone and large abdominal vessels. The flexura lienalis, adjacent descending colon, pancreas and lower duodenum were strikingly compressed forward and adhered to the tumor, but in these structures metastatic tumors were not found. Unexpectedly the left kidney seemed to be intact in appearence, but it adhered so tightly to the tumor at its upper pole, that the adhesion could hardly be dissected. The left adrenal was not found. Further, the tumor twined around the vertebra and large abdominal vessels, adhering to them, but the right kidney and adrenal were intact. In consequence, it was necessary to perform nephrectomy on the left side for total removal of the tumor, so we ligated and cut the left renal vessels and ureter. However, during further dissection of the tumor the infant became serious and died.

PATHOLOGICAL EXAMINATION : We missed an opportunity to perform a systematic autopsy. Therefore, any metastatic alteration could not be proved.

In hematoxylin-eosin preparation of the tumor, following findings were evidenced. Atrophied adrenal cortex was found in a part of the tumor (Fig. 2).

The tumor contained numerous blood vessels and scanty fibrous stroma. The cells of the tumor were round in type, as large as small lymphocytes in size, and the nuclei were spherical and rich in chromatin. The cytoplasms were scanty, clear and stained poorly. These cells resembled sympathogonia.

The most of them were arranged irregularly and disorderly, but some of them were arranged in packets. Wright had called attention to a tendency to form pseudorosettes (Fig. 5) as a characteristic of the embryonal sympathetic nervous tissues. In the Bielshowsky's preparation, intercellular neurofibrils were not proved. The left kidney was perfectly intact. Histopatholgical diagnosis was a sympathicogonioma of the adrenal medulla.

DISCUSSION

The tumor in our case was preoperatively supposed to be a renal tumor according to clinical findings and roentgenologic pictures, but it was revealed to be an adrenal one at operation. And development of the tumor was rapid enough to suggest a malignant tumor, though any metastatic tumor was confirmed neither clinically nor pathologically.

Moreover, the tumor showed following pathological features, which are chara-

cteristic of sympathicogonioma of the adrenal medulla ; namely,

(1) Constituent cells resemble sympathogonia in that they are as large as small lymphocytes and have a spherical or sometimes pyriform dense nucleus and scanty cytoplasm. (2) The tumor has scanty fibrous stroma and abundant blood vessels. (3) The cells have a tendency to form pseudorosettes, which are common in embryonal sympathetic nervous tissues. (4) Intercellular neurofibrils are not found in the Bielshowsky's preparation. The neurofibrils develop less intensely in sympathicogonioma than in sympathicoblastoma. (5) Atrophied adrenal cortex is found in a part of the tumor.

Sympathicoblastoma is composed of sympathoblasts, which are larger in size, have less dense nucleus and contain more cytoplasms than sympathogonia.

Thus the tumor in our case is undoubtedly a sympathicogonioma of the adrenal medulla.

SUMMARY

A case of sympathicogonioma arising from the left adrenal medulla has been reported. The case, 9 months old infant, presented the signs and symptoms of intestinal obstruction by the compression of a large tumor in left hypochondrium. There was no clinically demonstrable metastasis. The tumor was preoperatively diagnosed as a renal tumor, but it was revealed histologically to be a sympathicogonioma of left adrenal medulla.

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和文抄録

副腎髄質交感神経形成細胞腫の一例

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副腎髄質に原発した交感神経形成細胞腫の一例を報告した．本症例は生後9カ月の女児で，頑固な便秘及び嘔吐を主訴として，小児科に入院したが，左季肋下部に超手拳大，弾生硬，粗大顆粒状の腫瘤を触知し，レ線的に脾彎曲部より下行結腸上部に亘り腫瘤の圧迫に基く通過障害を認め，排泄性腎盂撮影では左側腎盂及び尿管像を全く証明し得なかつた上にアドレナリン注射後腫瘤容積の縮小を認めなかつた．依つて外科に転じたが，その翌日より嘔吐更に頻回となり，遂には糞様吐物を認めるにいたり，開腹術を敢行した処，手術は不幸不成功に終つたが，本症例に認められた腫瘤は吾々が術前考えた様な肝臓腫瘍ではなくて，副腎より発生した腫瘍であることが明かとなつた．即ち，腫瘤は後腹膜部に於て左腎上方より発生し，左腎前面を全く被い且夫れを後下方へ圧排して，その上極部に於て密に癒着し，脊柱，腹部大動脈，下空静脈を越え，右端は右腎に近接していた．更に脾彎曲部，下行結腸上部，膵及び十二指腸を夫々強く前方へ圧し，これらとも密に癒着していた．

併しながら肝臓，腸間膜淋巴腺及び上記後腹膜諸臓器には転移を思わせる様な変化は認められず，両側腎臓，右副腎も外見上正常であつたが，左副腎はその存すべき部位に於て発見することが出来なかつた．

病理組織学的に本腫瘍は次の如き交感神経形成細胞腫と考えるに足る所見を呈していた．

1) 腫瘍組織は少量の線維性間質による胞巣状構造を示し，血管に富んでいた．2) 腫瘍細胞は交感神経形成細胞に酷似し，略々淋巴球大，円形の細胞で，核質に富む球状の核並に僅少の原形質を有していた．3) 斯る細胞の大部分は不正雑然と配列していたが，一部の細胞群は所謂花冠状配列を示していた．このような花冠状配列像は胎生期の交感神経組織に認められるが又交感神経形成細胞腫の特異像である．4) 鍍銀標本では細胞間に無髄神経線維を証明できなかつた．交感神経形成細胞腫に於ては，その構成細胞である交感神経形成細胞が最も未分化である故に，神経線維形成は甚だ僅かであるか又は全然それを併わない．従つて，本症例の腫瘍が交感神経形成細胞腫であると考えられるが，更に腫瘍組織の一部に圧迫萎縮した副腎髄質に原発した交感神経形成細胞腫と云える．

Fig. 1. Schematic illustration of the tumor.

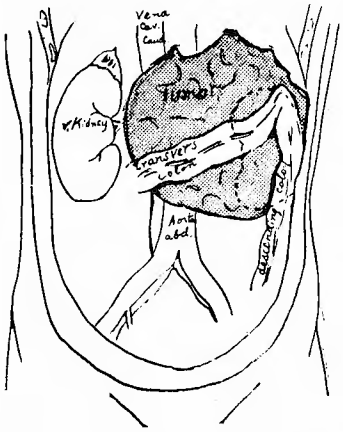

Fig. 2. Photomicrograph of the tumor.
A rim of adrenal cortex is shown in a part of tumor.

Fig. 3. Photomicrograph of the tumor.
(enlarged by 160)

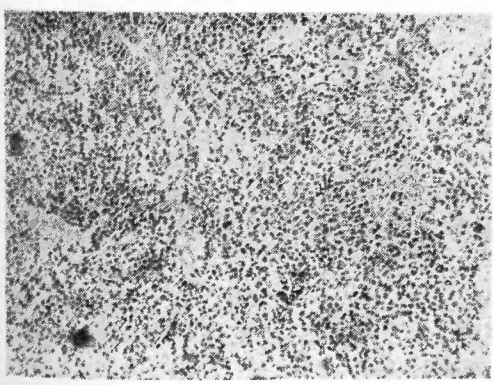

Fig. 4. Photomicrograph of the tumor.
(enlarged by 640)

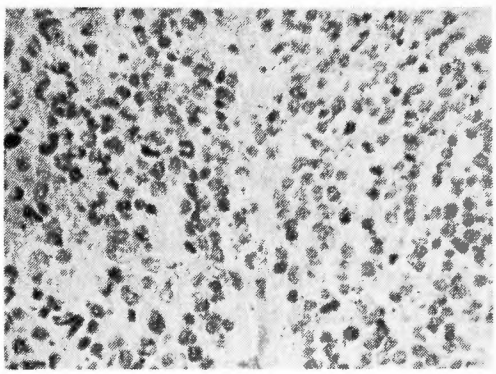

Fig. 5. Photomicrograph of the tumor.
Tendency to form pseudorosettes is indicated.