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<th>SYMPATHICOGONIOMA OF THE ADRENAL MEDULLA: REPORT OF A CASE</th>
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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 sympathetic 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 occurring 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 admission, 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 externally 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 xyphoides, 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 splenomegaly 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 appearance.

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 membrane. 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 appearance, 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. Histopathological 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-
characteristic 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.

REFERENCES

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か月の女児で、頭部の発育および嘔吐を主訴として、小児科に入院したが、左季節下部に手触り小発疹、亜発疹、熱で不退の腫瘍を触知し、レ線上の腫瘍部より下部を維持上部に及ぼす腫瘍の圧迫に基づく通気障害を認め、発祥性腫瘍を除く腫瘍性腫瘍を全で確認した上にアドレナリン注射後腫瘍の縮小を認めた。したがって外科的に転じが、その翌日より嘔吐更に厳重となり、遂には腫瘍を認めるにいたり、開胸術を施行したと、手術は不確不成功に終了し、本症例に認められた腫瘍は他と異なって特異な形態を示さなかった。副腎より発生した腫瘍であることが考えられた。即ち、腫瘍は後腹膜部に於って前方より発生し、左腎前面を全く覆い、腹膜を後方へ圧排して、その上脇部に於て密に発育し、脛柱、腹部大動脈、下肢脈を越え、右側は右脛に近接していた。実に腫瘍部より下部を維持上部に及び十二指腸を介して強く前方へ圧迫し、これらとも密に発育していた。

併しながら肝臓、腸間膜淋巴節及び上記後腹膜腫瘍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ったが、左副腎はその存在すべき部位に於いて発見することができなかった。

病理組織学的におきて、本腫瘍は次の如き変態神経形成細胞腫と考えるに足ると所を示していた。

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) 神経細胞は交感神経形成細胞に類似し、略々横大管大、円形の細胞で、核質に富む球状の核並に稀少の原形質を有していた。3) 基部細胞の一部は不正形且つ配列していたが、一部の細胞核は所謂花冠状配列を示していた。このような花冠状配列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) 腫瘍標本では組織間に線維性間質を認めることができなかった。交感神経形成細胞腫に於て、その構成細胞である交感神経細胞が最も未分化である故に、神経細胞形成は極度著しくあるか又は完全それを伴わない、従って、本症例の腫瘍は交感神経形成細胞腫であると考えられるが、実に腫瘍組織の一部に圧迫衰弱した副腎髄質に原発した交感神経形成細胞腫と言える。
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.
(enumered by 640)

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