

EXTRAMEDULLARY HEMOPOIETIC MASS COMPRESSING SPINAL CORD IN POSTERIOR MEDIASTINUM

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Extramedullary hemopoietic mass compressing spinal cord in posterior mediastinum

A case of posterior mediastinal mass diagnosed extramedullary hemopoiesis was presented. The patient had hemolytic anemia and spinal cord compression.

CASE REPORT

A 43-year-old man with chronic anemia had pain on his back and left costal margin, paraparesis, generalized weakness. Four years ago the patient had been admitted to Department of Hematology with hemolytic anemia, mild urinary retention, poor rectal sphincter tone, weakness of both lower extremities with spastic paresis, reduced reflexes and hypoesthesia at costal margin. His chest and skull X-ray had been normal. He had refused the myelography and he had left the hospital. Four years later he readmitted to Department of Thoracic Surgery with a right sided lobulary mediastinal mass.

On physical examination the patient was extremely pale and his scleras were subicteric. His liver edge was palpable 5 cm below the right costal margin and his spleen was 8 cm below the left. The former neurological signs were progressed. A moderate systolic murmur was heard on apex. The RBC count was 2.4 million, hemoglobin 7.9 gr. At lateral chest roentgenogram the posterior mediastinal mass was between the levels of T-6 and T-3 vertebral bodies and intervertebral spaces were irregular. At myelogram there was an obstruction at level T-6.

After the transfusion of 5 units of blood, the neurosurgeons operated on decompression laminectomy and the epidural mass extending from T-6 to T-9 into

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both paravertebral regions was removed. After the operation the neurological signs were disappeared. Then right posterolateral thoracotomy was performed and a lobular, 8x8x3 cm brownish granular tumor was extirpated. The histopathological diagnosis was reported as extramedullary hemopoiesis. After the postoperative recovery the patient was transferred to hematological clinic for the treatment of hemolytic anemia.

DISCUSSION

Extramedullary hemopoiesis in posterior mediastinum develops by extrusion of hyperplastic marrow through the thinned trabeculae in the proximal ends of the ribs. Other origins may be embryonic rests or lymph nodes. EMH is most frequently encountered in posterior mediastinum and lesser in posterior pelvic region (2,3,4). It can also cause spinal compression (1). It must be differentiated from neurogenic tumors, spinal tumors and tuberculous abscesses. In posterior mediastinal masses accompanied with chronic anemia EMH should be suspected. In this case radical removal of the mediastinal mass and decompression of the spine followed by radiation therapy is essential

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ANEURYSMAL BONE CYST (x)

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Summary: Four patients presenting with aneurysmal bone cyst were introduced. the first patient was 16 year old with a cyst of the humerus. The second was very interesting, but his cyst was diagnosed initially as a fibrous dysplasia. The Third patient was a twenty year old female. Her right fibula was involved, the cyst was assumed as a giant cell tumor at the beginning. In the forth patient right femur was affected and he was a 20 year old private. (Soldier) All the patients were treated surgically.

Aneurysmal bone cyst, described by Jaffe and Lichtenstein in 1942, is an-un-common benign bone lesion. Having unknown etiology, it is initially confused with malignant tumor (1,2,3,4,5,6,7,8,9,). Usually it is seen in adolescents and young adults with history of trauma, pain and swelling. When it is near the joint, it limits the movements and neurologic findings are seen it lies in vertebrae (3,4,7,8). Although it is most commonly observed in second and third decade, cases are seen in childhood and at the age of forty (3,4,5,8). The ratio of occurrence in females is two to one in reference to males (1,4,8). It presents 1 % of all bone tumors and 2 % of benign bone tumors. About its 75 % is seen in-femur, innominate bone and vertebrae. At least 50 % is located in long bone metaphysis, but it appears in all bones.

Histologically being separated by connective tissue septums, it is a lesion which has blood-filled spaces, bone trabeculations, osteoid tissues and giant osteoklast cells (2,3,4,5,7). It is one of those several disorders which gets its name from radiological appearances (3,5,8). Aneurysmal bone cysts are-purely osteolytic. Their borders are well-defined and they have sclerotic endosteal margin. The lesion is expansile and at the beginning excentric (2,3,4,5,6,7). The bone cortex may be thin or sometimes may be destroyed because of the progressive expansion (4). Usually, it develops gradually but sometimes it may be rapidly progressive ballooning bone and gets its typical appearance in a period of 6 weeks to 3 months

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(3,5,8). Frequently having the form of soap bubble, it makes a fusiform expansion when placed in the long bone shaft. The transition zone is generally narrow, sometimes the lesion develops rapidly and shows a wide transition zone with a soft tissue swelling. Therefore it is frequently mistaken for malignancy (5,8). If a fracture occurs in the lesion area, a periost reaction develops (4,7,9). When it is in vertebrae, it is mostly seen at the dorsal and lumbar level or in vertebral accessories. In this location, beside the lytic and expansile features, erosions may be seen in vertebra of costa (5,8). It causes asymmetry when lies on face bones (3). Hypervascularity is noticed near tumor by angiography (4,5).

Differential diagnosis: With a rapid growing it may be mistaken for a malignant tumor such as osteosarcoma and giant cell tumor (3,5,7,8,9). The giant cell tumor is often seen after the epiphyseal fusion and observed around the knee. Rapid development, excessive bone destruction, new bone formation and wide transition zone in osteosarcoma are helpful for differential diagnosis. Angiosarcoma can be diagnosed by angiography (3,5). Although it is confused with plasmacytoma and metastases in vertebrae, the age of the patient is an important clue (5). Fibrous dysplasia and hemophylic pseudotumor should be considered in differential diagnosis (5).

Treatment: In the treatment curettage and radiotherapy is used. Sometimes, even a simple biopsy can subside the growth. The prognosis is completely benign (1,2,3,4,5,6,7,8,9).

Case reports:

Case 1. The arm of a 16 year old girl was broken when she was at the age of two. It was put in plaster by a village medicine man. The swelling did not disappear completely, but developed in the course of time with a rapid speed in the following year. With its swelling which limited the movements of the elbow, a slight pain and itching was felt occasionally at this region. In the inspection physical retardation and weakness was observed. In the physical examination there was a firm, painless mass having venous appearances on it measuring 23x24-cm. in the upper elbow joint of the right arm. Because of this mass they had to have a zip fastener on the right arm (Fig. 1,2,3). X-ray showed a bone lesion which measured 20x22 cm. in the distal metaphysis of humerus covering the elbow joint. It had lytic, expansile well-defined contour, sclerotic endosteal margin septas and partly intact bone zones. Extreme expansion made the bone cortex get thinner and partly destroyed (Fig. 4,5,6). The mass was removed with an operation and the left fibula was put in the defective field. She was put in abduction plaster and was discharged to be examined again three months later. Measuring macroscopically 13x15x20 cm., it had a yellow wall about two centimeters thick and full of hyperemic and blood-filled spaces. It was a partly hard bone. The blood-filled spaces separated by connective tissue septas (Fig. 7). Histologically a lot of multinuclear



Figur - 1

giant cells were observed with bone and connective tissue septas, osteoclastic and osteoblastic proliferations. It was considered an aneurysmal bone cyst. The patient was able to use her right arm when she came for a check up after six months. It was noticed in the radiogram that the medullar nails and fibular graft were in the desired form (Fig. 8). After the removal of the medullar nails it was observed that the bone graft was sufficient and there was no difficult in using her arm. The patient who is under the care of our staff for the past three years has no problem now (Fig. 9).

Case 2. A 22 year old man came to us with complaints of pain and swelling on his chest for two years. He said had did not remember a remarkable trauma, but felt that the swelling got larger and harder. In the physical examination a hard and painful mass was found at the level of third, fourth and fifth costa. It was in the shape of hemisphere with a smooth surface and five centimeters in diameter at the level of intercostal spaces. X-Ray showed a lesion which had septas, a fusiform extension and lucent. It made the bone cortex thinner measuring 5x10 cm. in the corpus sterni (Fig. 10). The macroscopic appearance of the cystic mass which had a highly bleeding operation and was removed by mediosternotomy was atypic, hyperemic and irregular. In its histology there were osteoclastic and osteoblastic giant cells and connective tissue septas. Angiomatous differences were observed between fibrosists an fibroblasts (elements of connective tissues) that formed the stroma. With the finding of multinuclear giant cells, it resembled giant cell tumor, but the absence of malignancy findings made us think them as aneurysmal



Figur - 2

bone cyst. It was observed that the patient had a very comfortable and healthy life for more than four years.

Case 3. A twenty year old woman presented with complaints of pain and swelling at the right ankle. She said hat the pain and swelling gradually increased recently. When we questioned her about trauma, she said that especcially her right ankle was often distorted. In the physical examination a painful swelling measuring 7x10 cm. was observed at the outer malleol of the right ankle. The mass was smooth. X-Ray showed a lesion measuring 6x10 cm. in the distal metaphysis of the right fibula. It had lytic, expansil, a partly destroyed and weakened bone cortex, septas with various appearances and intact bone particles (Fig. 11,12). Middle fibula was fixed at tibia with two screws after removing the cyst by 1/3 right distal fibula resection so that right ankle joint could work appropriately (Fig. 13,14). The patient was put in long leg plaster and discharged for a return after 1.5 month. In the histological exam it was concluded as aneurysmal bone cyst. We have ob-



Figur - 3

served that the patient has been walking easily for three years and her ankle has its normal function.

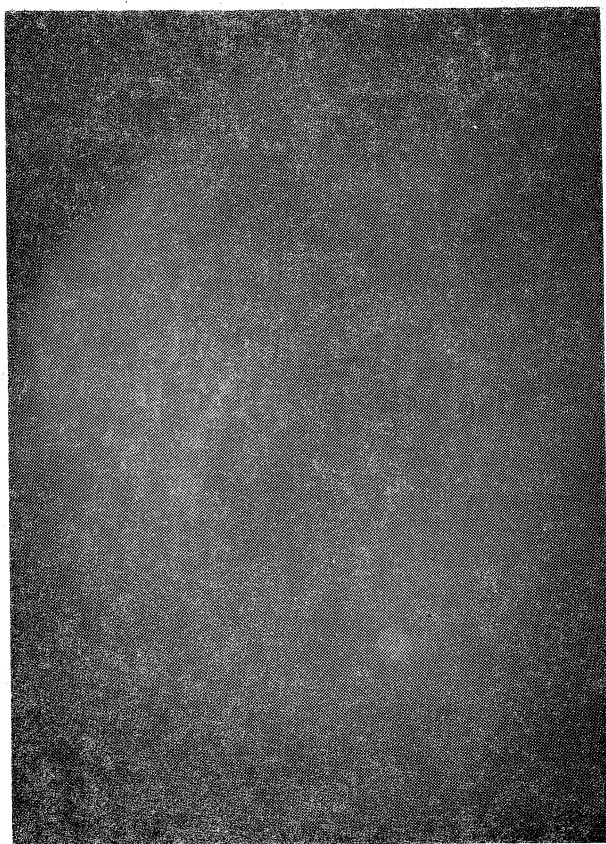
Case 4. A 20 year old soldier complained of a sudden pain at his right thigh while making exercises and not being able to walk on it. He had no trouble before. X-Ray showed a pathologic fracture in right femur intertrochanteric area because of a lytic defective lesion with septa. It was thought that medial cortex was displaced into the soft tissue after being fractured (Fig. 15,16). Noticing a pathologic fracture at the operation, with a lesion curettage the iliac graft and medullar nailing was done (Smith Peterson Nail). He was discharged to be checked again after three months. After histologic exam it was certainly thought to be an aneurysmal bone cyst. But we had no chance of checking the patient after he was discharged from the army.

Discussion: With its unknown etiology, aneurysmal bone cyst is an entity receiving its name from the X-ray appearance. But as trauma is often seen in the



Figur - 4

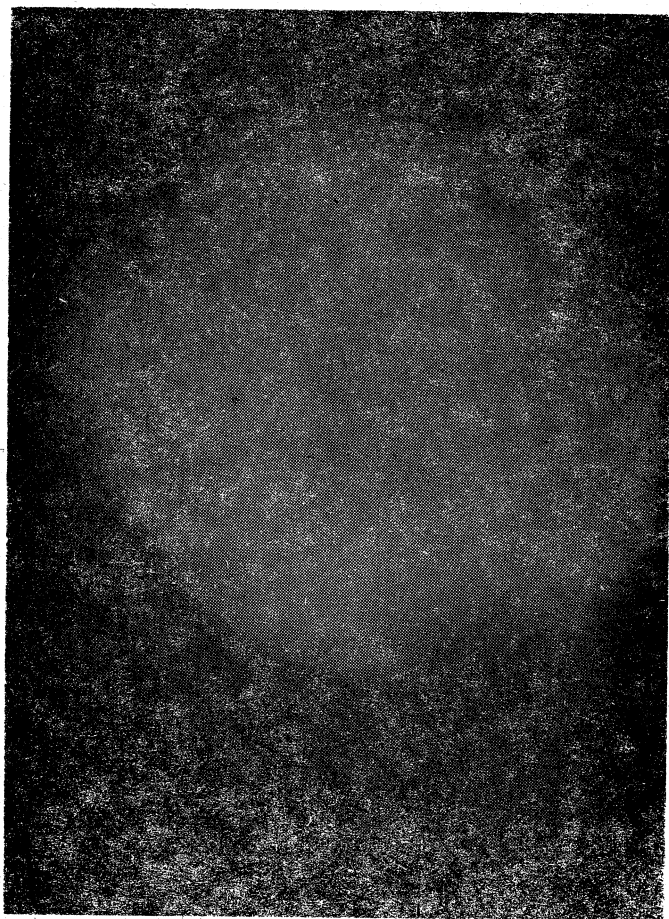
history, its considered likely to be related with it (1,2,3,4,5,6,7,8,9). There was history of trauma in our three cases. In the first, trauma was recognized but no certain evidence of its history. In the second, although undoubtedly it is not a special trauma. The fact that young athletes have trauma while playing basketball is certain. In the third case, the history of frequent distortion of her right ankle clearly points to trauma. Levine at al claim that bleeding of arterio-venous shunt has been so stronger and closer that at cystic mass is formed in the one. If the bleeding occurs under periost, it may be damaged so that subperiosteal cystic lesion develops. The some authors emphasize that the capsuled hematoma formed after trauma is pseudotumor. According to some other claims the traumatic aneurysmal bone cyst has been developed out of the bone. According to these opinions the type developed in the bone is considered likely to be a cystic or telangiectatic giant cell tumor. The development pattern of two type is different but at the end they have the same radiologic and histologic characteristics. The ones that developed in the bone enlarge the bone cortex whereas the ones that develop out of the bone



Figur - 5

give a radiologic appearance resembling soft tissue tumors. The relationship with trauma is not yet completely clear. Because it can only be recognized at a few patients. Lesion is lytic but its margins are well-defined and sclerotic (3,5,7,8). In spite of the rapid growth and ballooning, a slight destruction on the surrounding bone and the narrowness of the transition zone (2-3 mm.) show that lesion is benign (5,8). Although it may be found at every bone, it is mostly seen in the long bone metaphysis, innominate bone and vertebrae (3,5,7,8). In our cases it was located at lesion distal metaphysis of the humerus and fibula, upper femur metaphysis and in sternum which is rarely involved. As it is seen at all bone tumors, importance of trauma aneurysmal bone cyst may arise from preexisting bone lesions, such as chondroblastoma, giant cell tumor, chondromyxoid fibroma, xantoma or osteosarcoma (3).

Therefore it must be kept in mind that it may be associated with benign or malignant bone lesion apart from trauma (3,7).



Figur - 6

Curettage or biopsy may be curable (2,3,5,7,8). If the lesion is on an unimportant bone, the bone may be removed totally (8). Radiotherapy is used for treatment, but it is feared that it may lead to sarcomatous changes (7). Because the operation may be dangerous and lead to bleeding. Radiotherapy is usually applied on vertebral lesions (8). Besides the other specification of radiotherapy emphasises that this lesion shows at least a pseudomalign peculiarity (8). Although curettage and radiotherapy may be curable, there may be recurrence. Therefore they are not completely safe and there may be a risk even though the chance is so small.

Operation is performed in all our four cases. There has been no problem in the first case for three years. The second case has been living for four years. The third case has been using her leg very well for three years. The fourth patient could not be studied as he left the army. There was the obvious trauma history in three of these, but it was unclear only in the fourth one.

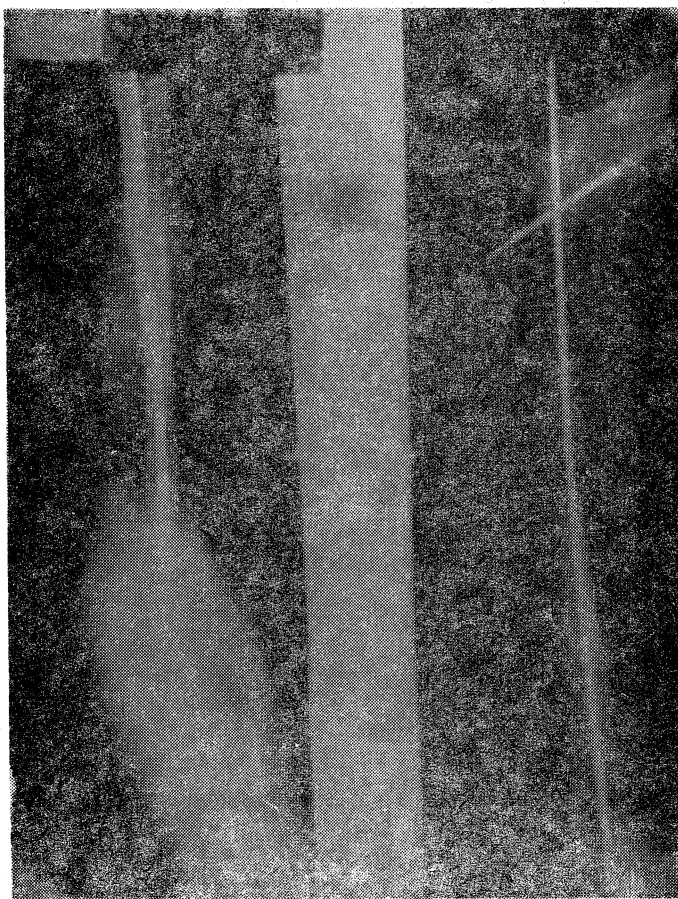


Figur - 7

With this research we have understood that trauma has a very important influence in the development of the aneurysmal bone cyst.

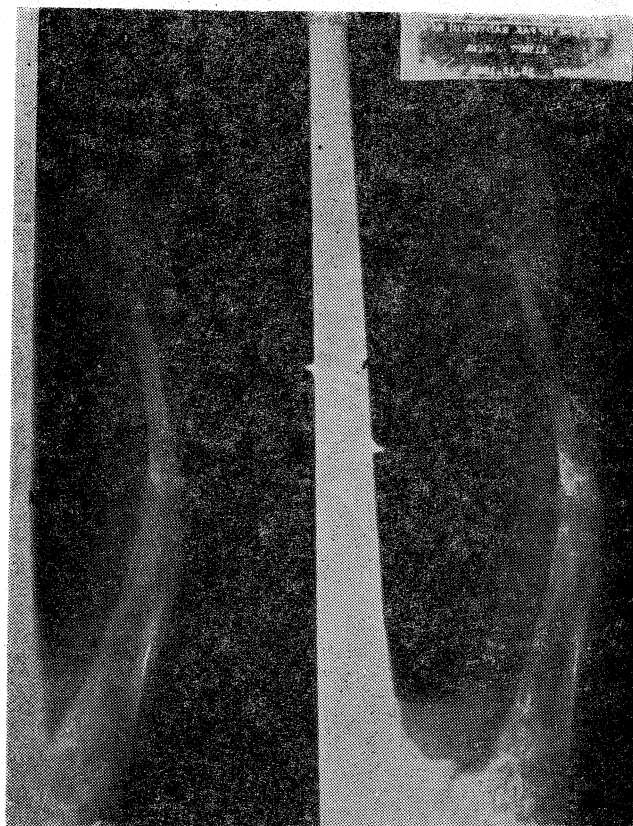
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Figur - 8

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Figur - 9



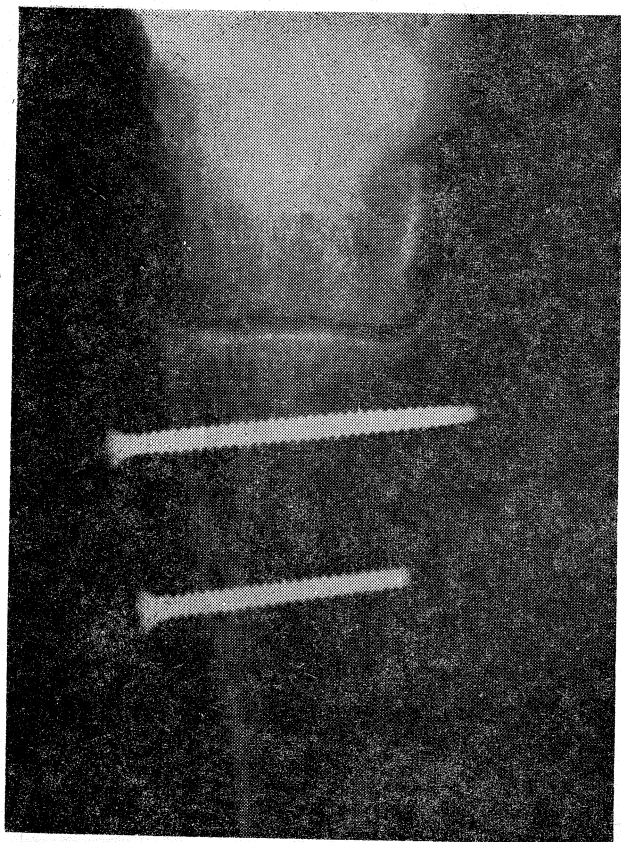
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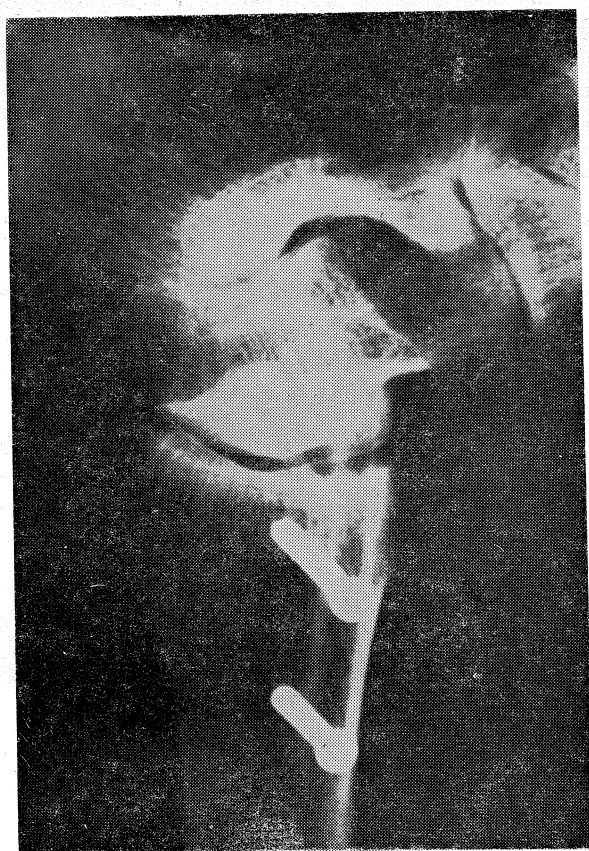
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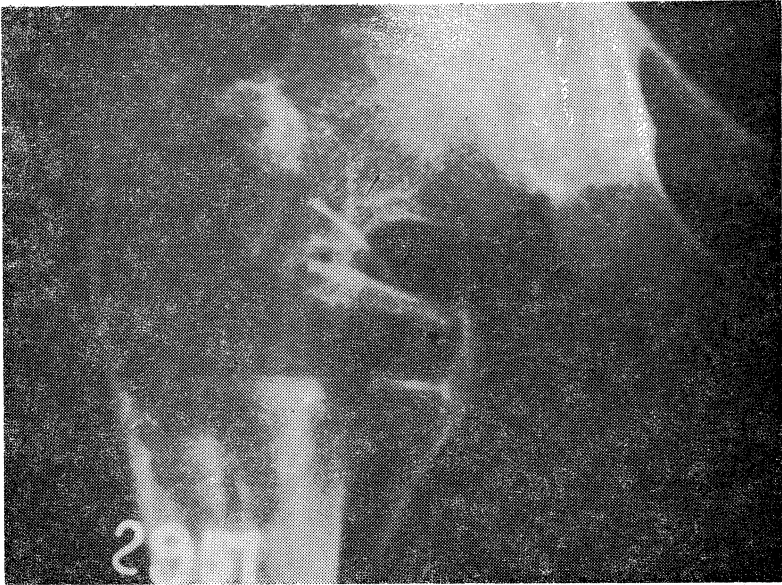
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Figur - 13



Figur - 14



Figur - 15



Figur - 16