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**HEMANGIOENDOTHELIOSARCOMA OF THE ILEUM WITH  
METASTASES TO THE RIGHT AURICLE, RUPTURE OF  
THE HEART, AND HEMOPERICARDIUM AT LEAST  
10 DAYS BEFORE DEATH: A CASE REPORT \***

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A CAREFUL review of the available literature fails to reveal a case similar to the one presented in this paper. Because of the exceeding rarity of hemangio-endotheliosarcoma, and the unique complication of rupture of the heart due to metastatic involvement of the right auricle, this case is considered worthy of reporting.

*Incidence:* Sarcoma of any kind of the small intestine is rare,<sup>2</sup> and hemangio-endothelioma itself is rare among the sarcomas.<sup>3</sup> "In the New England Deaconess and Palmer Memorial Hospitals from 1927 to 1939, Medinger was able to find only 10 malignant tumors of the small intestine at autopsy and in the surgical material studied, there were 12 instances. In other words, in 42,456 examinations, he found 22 instances of malignancy of the small intestine and of these only six were sarcomata."<sup>2</sup> Maxwell<sup>1</sup> states that all malignant tumors of the small intestine form only 3 per cent of the malignant tumors found throughout the intestinal tract.

*Classification:* Legg and Fitch<sup>4</sup> indicate that vascular tumors present a wide histogenetic range which, in order of activity, may be grouped as follows:

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1. The typically benign angioma.
2. The multicentric benign angioma.
3. The cellular capillary angioma which occurs in children and is locally invasive (hemangioma hypertrophicum).
4. The locally invasive, plexiform angioma, which may invade muscle and even bone.
5. The genuine malignant angioma or true hemangioendothelioma.

*Site of Origin:* Stein<sup>6</sup> states that sarcoma is more frequently found in the small bowel than in the large bowel, and that it occurs more often in the ileum than in either the duodenum or the jejunum. According to Kastl,<sup>3</sup> hemangioendothelioma shows no special predilection for any single part of the body, having been reported in almost every organ.

*The Patient:* The majority of cases of sarcoma of the small bowel occur between the ages of 30 and 40, whereas carcinomas are more often found in patients from 40 to 60 years of age.<sup>6</sup> There is no marked sex preponderance.<sup>3</sup>

*Metastases:* As to hemangioendotheliomas, metastases occur as a rule by way of the blood stream, but occasionally by way of the lymphatics. The lungs are most frequently involved, with metastases to the liver, spleen, kidneys, pancreas, adrenals, bones, lymph nodes, and peritoneum reported.<sup>4</sup>

Stein states that metastases from malignant tumors of the small intestine occur very early in the disease.<sup>6</sup>

*Symptoms:* There are no characteristic symptoms associated with tumors of the small bowel. The most common symptomatology found in the majority of cases is that of intermittent partial obstruction.<sup>6</sup>

*Prognosis:* The prognosis for malignant tumors of the small intestine, whether operable or inoperable, is poor.<sup>1, 6</sup>

#### CASE REPORT

A 71 year old white female, wife of a local pastor, entered the hospital\* February 2, 1953. Her chief complaints on admission were marked weakness, moderate dyspnea on exertion, and belching of approximately one month's duration. A careful inventory of systems was otherwise unrevealing. There had been no evident bleeding from any of the body orifices, and no melena.

*Past History:* Noncontributory except for "peritonitis and locked bowels" 40 years before, which had been treated medically.

*Family History:* Noncontributory except for a sister who had had "cancer of the colon" in 1952, at the age of 58, and had been operated on for this condition.

*Physical Examination:* The patient was a lively, intelligent, coöperative lady, well preserved for her years, who walked easily about her hospital room on admission. Physical examination revealed only two significant findings: her skin was very pale, and the liver was symmetrically enlarged to a distance of four fingerbreadths below the right subcostal margin. The heart tones were heard easily, the rhythm was regular, and there were no murmurs. The blood pressure was 120/80 mm. of Hg.

*Laboratory Examination:* Stool examinations were repeatedly made. Melena was never present, but a markedly positive immediate reaction to the benzidine test for occult blood was demonstrated each time.

\* Perry County Memorial Hospital, Perryville, Missouri.

*Blood Studies* (on admission): Red blood cells, 2,750,000; white blood cells, 7,500; hemoglobin, below 47 per cent; hematocrit, 22 per cent. Differential: basophils, 4; stabs, 6; segmented, 56; lymphocytes, 34. Bleeding time, 1 minute; coagulation time (capillary tube): 4 minutes 20 seconds; platelets: 205,500 per cubic millimeter; blood type, A; Rh negative. Bone marrow aspiration revealed a mild normoblastic hyperplasia; peripheral blood smears marked hypochromia.

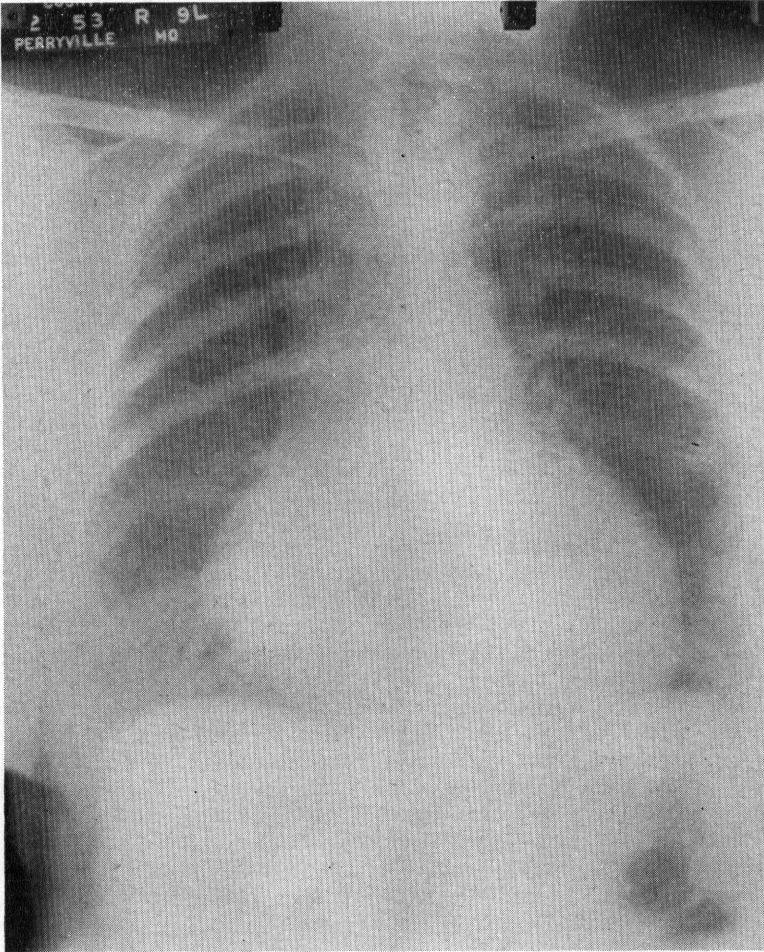


FIG. 1. Chest x-ray performed 10 days before death reveals marked enlargement of the cardiac silhouette to both the right and the left. The enlargement was due entirely to a massive hemopericardium.

*Urinalysis* was essentially negative.

*Blood Chemistries*: Nonprotein nitrogen, 34 mg. per cent; fasting blood sugar, 86 mg. per cent; prothrombin time, 25 seconds.

*X-Ray Studies*: On February 4, two days after admission, fluoroscopic and radiographic examination of the chest (see figure 1) revealed marked cardiac enlargement to both the right and left. There was no pulmonary edema or fluid. (Note that this was 10 days before death.)

A gall-bladder series on the same date indicated a nonfunctioning gall-bladder, with no evidence of opaque calculi. There was evidence to indicate marked hepatomegaly.

A complete upper and lower gastrointestinal series, including a small bowel study, revealed no evidence of pathology in the gastrointestinal tract.

*Electrocardiogram:* A routine 12-lead electrocardiogram on February 3 revealed an intermediate position, low voltage in all leads, and no S-T segment or T wave abnormalities.

*Clinical Course:* The patient had no other complaints, except for the original symptoms of weakness and dyspnea, for the first week of hospitalization. At this time her laboratory work-up had been completed and it had been decided to get her in condition for an exploratory laparotomy.

On February 8 she complained of increasing weakness, and 500 c.c. of whole blood were administered. She stated she felt better after the transfusion.

The next day she complained of more weakness, dyspnea and right upper quadrant pain. There was no orthopnea. The blood pressure was 120/96 mm. of Hg, the heart rate 110 and regular. There were a few crepitant râles at both bases. The liver was definitely larger and very tender, and was palpated 5 to 6 fingerbreadths down. The patient was then given 1.2 mg. digitoxin in two doses, and oxygen was administered.

The following five days were characterized by a continued downward course. Despite more digitoxin, oxygen and repeated small transfusions, the patient gradually became weaker and more dyspneic. She suffered continuous pain from her enlarging liver.

The patient died on February 14, 1953, 12 days after admission.

#### NECROPSY REPORT \*

*Gross Findings:* A tumor, 1 cm. in diameter and 4 mm. thick, was found in the terminal ileum. The tumor had eroded through the serosa. The mucosal surface presented the appearance of a small red velvety button.

One small tumor was found in the mesentery of the small bowel, a similar tumor in the greater omentum.

The liver substance had been practically replaced by tumor. Round fleshy tumors were studded throughout the liver, varying from the size of a pea to that of an orange. The gall-bladder and other viscera were normal.

When the chest cavity was opened, the cause of the cardiac enlargement was immediately perceived. The pericardium was greatly distended and bluish in color. Incision of the pericardium released about 500 c.c. of nonclotted old blood. Before the blood had been completely evacuated from the pericardial cavity, and before any incision had been made into the heart, blood was seen pouring from a matchhead-sized perforation in the anterior wall of the right auricle.

Careful dissection of the heart revealed tumor involving various parts of the right auricle and the right auriculoventricular septum. There was no tumor elsewhere. Careful inspection of the coronary arteries revealed no evidence of thrombosis or occlusion. Serial sections of the ventricular myocardium revealed no evidence of infarction or tumor.

*Microscopic Findings:* † Microscopic studies of sections taken through the ileum, liver and right auricle revealed characteristics typical of hemangioendotheliosarcoma.

\* Autopsy was performed with the kind assistance of W. F. Utterman, M.D., Perryville, Missouri.

† Microscopic studies were performed by T. A. Lovinggood, M.D., Cape Girardeau, Missouri.

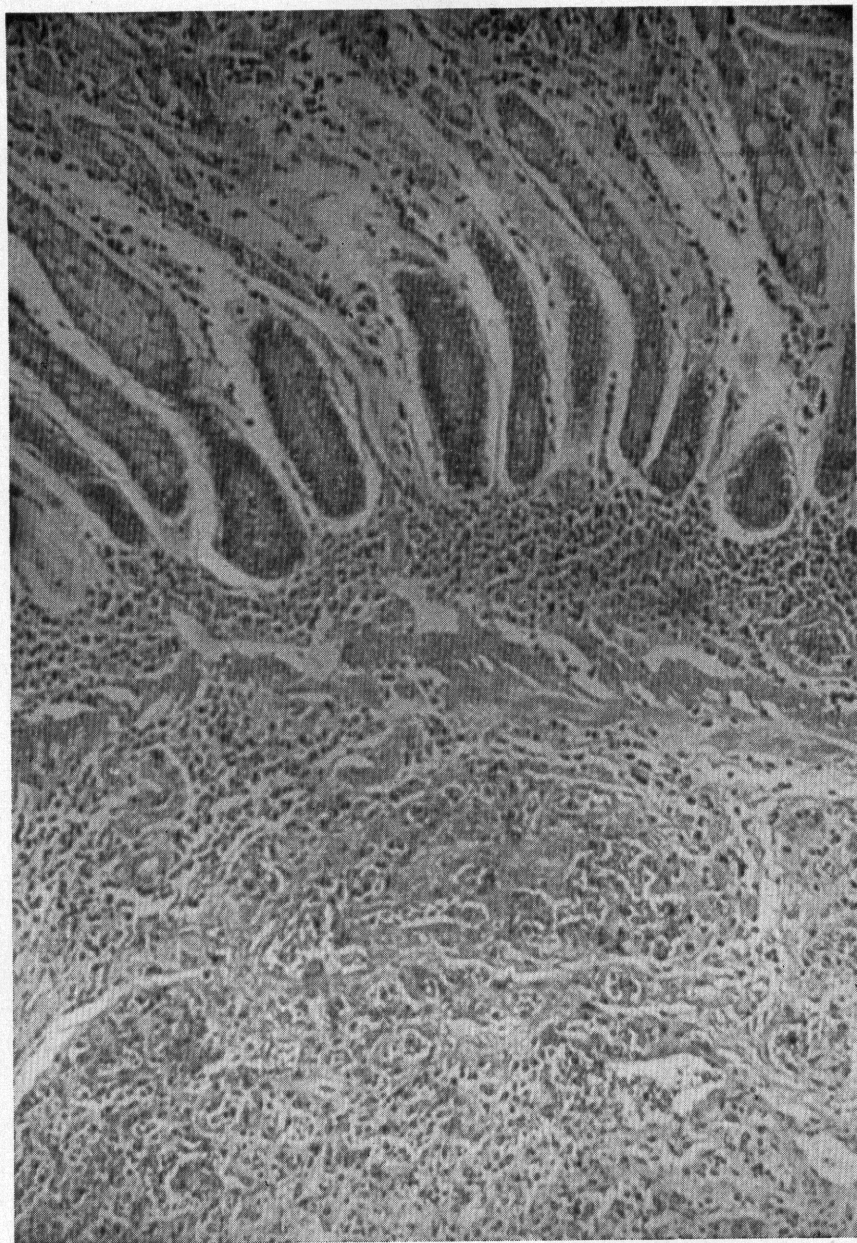


FIG. 2. Section taken through the primary tumor in the ileum. The tumor is seen to consist of numerous tiny capillary spaces lined by flattened endothelial cells.

A section (figure 2) taken through the previously mentioned button-shaped tumor of the terminal ileum revealed tumor in the mucosa and serosa. It was seen to be composed of numerous tiny capillary spaces lined by flattened endothelial cells. In this area only the formation and growth of capillary spaces were noted. There

was very little evidence of intraluminal proliferation, and many of the small spaces were filled with erythrocytes. There were some chronic inflammatory cells infiltrating at the periphery of the tumor and also through the interstices of the tumor itself. The tumor tended to be irregularly lobulated.

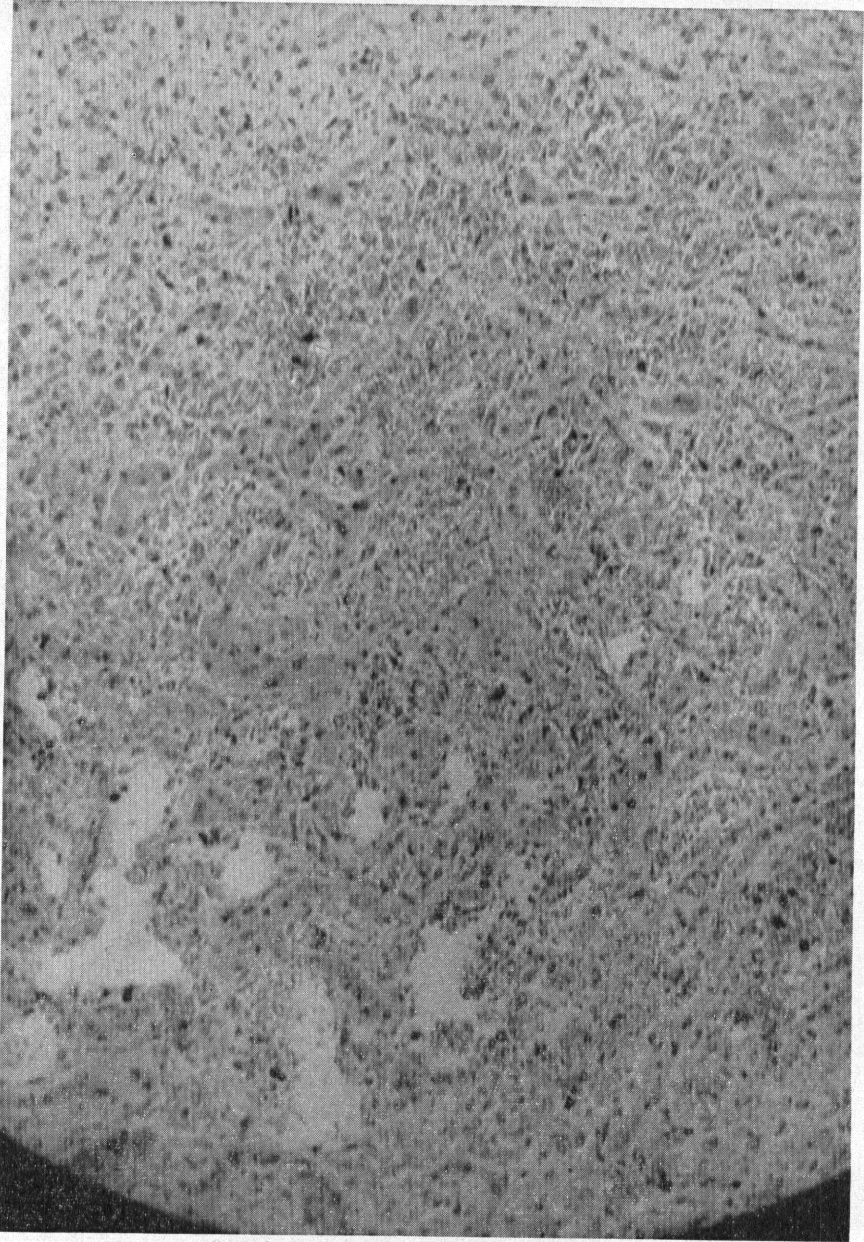


FIG. 3. Section through the liver with a portion of tumor included reveals the same pattern as in the primary tumor, but with more anaplasia of the endothelial cells.

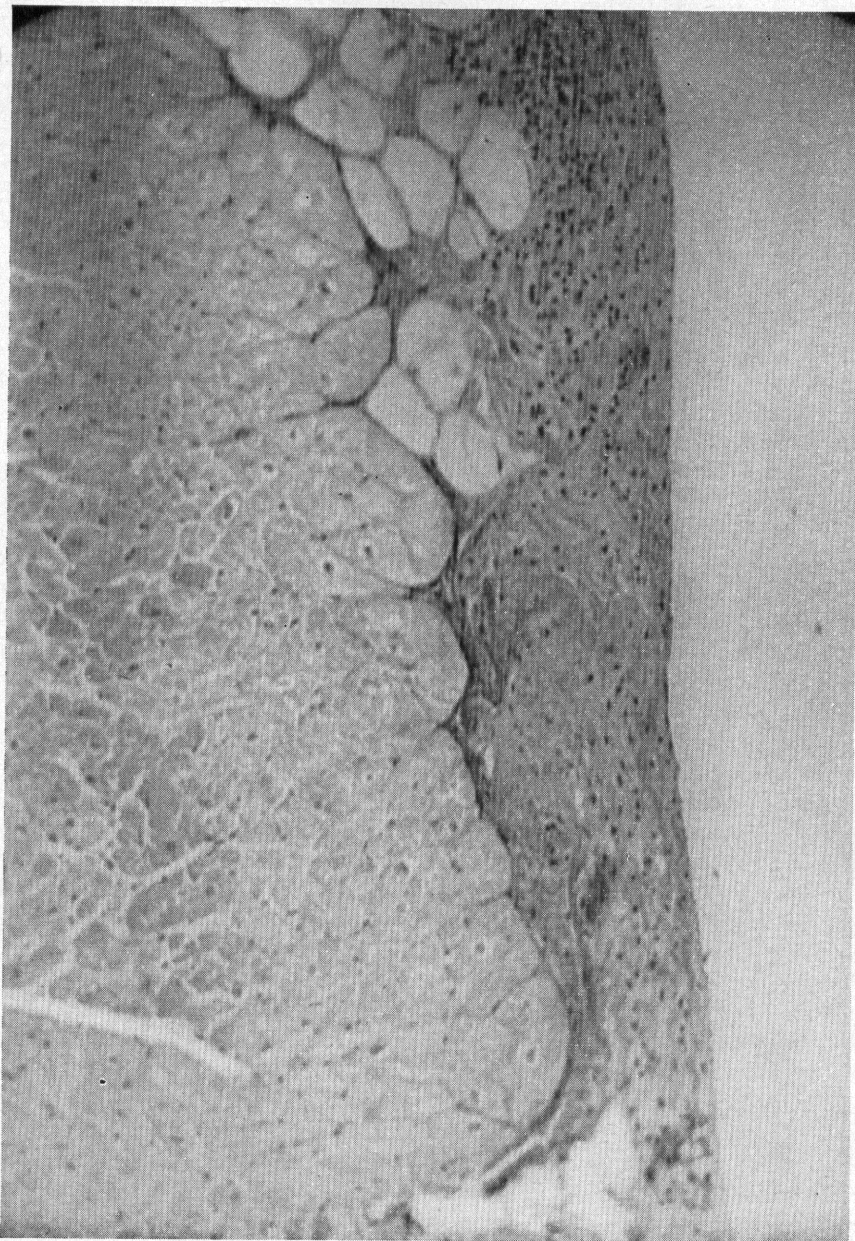


FIG. 4. Section through the right auricular myocardium reveals diffuse tumor growth. It was the spontaneous perforation of one of these metastases which was responsible for the hemopericardium.

A section taken through the liver with a portion of tumor included (figure 3) showed again the well formed capillary pattern, but in this instance there was more anaplasia of the endothelial cells and more intraluminal proliferation. In addition to this, there was quite a bit of disorderly growth of the capillaries which penetrated into the liver lobules. The tumor tended to be rounded and nodular in outline, as in the bowel. Again, erythrocytes were noted within the lumen of many of the capillaries.

A section through the myocardium (figure 4) revealed a diffuse tumor growth, made up again of capillaries proliferating in a very irregular manner. Anaplasia of the lining endothelial cells was not marked. There was some irregularity in many of the nuclei of the cells. The capillaries again contained a few erythrocytes.

#### COMMENT

This is a case of hemangioendotheliosarcoma of the ileum in a 71 year old woman which first manifested its presence in the patient by bleeding slowly and thus producing a severe anemia. Apparently the anemia had been present about one and one-half to two months before death, and in view of the natural history of this type of lesion it is probable that the original tumor had not been present much longer.

It is to be noted that this original tumor, which was to eventuate in death, was so small and so remote in the recesses of the body that all available laboratory and diagnostic aids could not demonstrate it. Only laparotomy would have been successful in demonstrating the lesion. In view of the huge, widespread metastases found at autopsy, it is doubtful that surgery at any time during the clinical course of illness could have been of benefit, even had the patient been operated on at the first vague signs of illness one month before.

A remarkable observation was the largeness of the tumor implants in the liver. The site of origin may be debated, but it seems logical that if the tumors in the liver were primary and the tumor of the ileum was an implant, then there should have been more than one implant. Furthermore, it would seem too great a coincidence to expect one implant to choose the most frequent site of sarcoma of the entire gastrointestinal tract. The most reasonable supposition is that the tumor originated in the terminal ileum and began to bleed slowly but steadily due to the necrotizing effect of proteolytic enzymes in the lumen of the ileum. It then invaded the portal system and metastasized to the liver. From there, tumor cells no doubt migrated through the hepatic venous system to the right side of the heart, where invasion of the auricular musculature took place. Why metastases were not found in the lungs is difficult to say.

Spontaneous rupture of the right auricle definitely had occurred at least 10 days—and probably longer—before death, at a time when the patient was fully ambulant and did not appear very ill. This is proved by the enlargement of the cardiac silhouette as seen on February 4, 1953. At this time a huge hemopericardium was already present. Inasmuch as rapid filling of the pericardium with blood under great pressure, as from a ruptured myocardial infarct, causes tamponade and sudden death, the enlargement of the pericardial sac and its distention with blood must necessarily have taken place over a relatively long period of time. The estimation of this period of time is strictly conjectural, but it is possible that rupture occurred more than a week before the x-ray was taken. Thus, rupture



of the heart with hemopericardium was present at least 10 days—and possibly as long as three weeks—before death.

#### SUMMARY

1. A report is presented of a case of hemangioendotheliosarcoma of the ileum with metastases to the liver and to the heart.

2. Evidence is presented which indicates that spontaneous rupture of the right auricle with massive hemopericardium had occurred at least 10 days—and possibly as long as three weeks—before death, while the patient was still ambulant and clinically only mildly ill.

3. A brief review of the available literature concerning this disease is presented.

4. The significance of various aspects of the case is discussed.

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