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PLASMOCYTOMA OF THE MIDDLE EAR

by

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PLASMOCYTOMA OF THE MIDDLE EAR

K. H. Goll and G. Rumpf

The authors describe an extramedullary mucosal plasmocytoma of the middle ear and mastoid mucosa, which was autotomized. The diagnosis has been secured in cytological, histological and postmortem findings. The plasma cell infiltration was confined to the mucosa. In the operational region there was no histological indication of an affection of the bones. The X-ray picture did not show any safe osteological changes in the remaining skeletal system either. There existed, however, all clinical symptoms of a diffuse plasmocytoma with corresponding changes in the bone marrow, paraproteinemia, proteinuria without Bence-Jones protein, anemia and protergent development up to the exitus due to pneumonia. Contrary to the opinion that extramedullary plasma cell tumors represent a pathological picture of their own, the authors hold that these plasmocytomas also belong to the category of diffuse myelomata, distinguishing themselves by a peculiar progressive form.

The study of the extramedullary mucosal plasmocytomas in the region of the ear, nose and throat presents an interesting contribution to the pathogenetic total concept of plasmacellular proliferations. The mucosal plasmocytoma of the middle ear or the mastoid mucosa as far as we know has only once been described in this relationship up to now [1]. In addition, another case was described together with a deformation of the ear. In the following article a case such as this is to be discussed.

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Patient B. M., age 83.

History: Pulmonary tuberculosis since age 29, at age 30 Fothergill's operation. Anemia for about 5 years and for a quarter of a year he has been hard of hearing in both ears. Since about a week ago pain in the left ear.

Hospitalized due to otitis media with mastoiditis on November 14, 1961.

Local findings: Left drum membrane: Bright red, thickened, protuberant. Retroauricularly reddened, mastoid pain.

Right drum membrane: No pathological findings.

In the hearing test from the right ear the patient could hear numbers spoken at a whisper from a distance of 4 m, and speech from a distance of 6 m. The corresponding values for the diseased ear amounted to 20 cm and 2.5 m, respectively. In the audiogram the acoustic perception in the right ear corresponded to the age, whereas on the diseased side the disturbance in the acoustic perception was accompanied by a bone conduction disturbance which amounted, on the average, to 30 Db.

In the X-rays that were taken according to Stenvers there were flat, limited upper pyramidal edges. The porous auditus internus was inconspicuous on both sides. The picture according to Schüller showed a conspicuous and well defined pneumatization on both sides. The individual cell trabeculae were clearly recognizable also on the diseased side which, in comparison to the normal side, clearly showed less radiopacity.

With the exception of the slight rhinitis there was no other finding in the region of the ear, nose and throat.

Other findings: Systolic murmur over the heart, blood pressure: RR 190/100. The liver was about 4 cm below the right rib arch,

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the spleen was palpable at the right rib arch. There were no lymphatic node swellings. Bilateral inguinal hernia, no skeletal pain, normal reflexes, no pathological findings in the other organs.

Laboratory findings: Blood sedimentation 135/158, hemoglobin 7.2 g/100 ml, erythrocytes 2.32 million, leukocytes 3400; differential blood picture: basophile 0, eosinophile 2, immature 0, stab-like 5, segment-like 59, lymphocytes 30, monocytes 4. Anisocytosis, poikilocytosis, ovalocytosis. Electrophoresis: albumin 42.0%, α_1 4.5%, α_2 5.8%, β 10.7%, γ 37.0%.

There was albumin in the urine, the Bence-Jones protein was especially negative. Due to the anemia a sternal puncture was made: the smear was cell rich, which was dominated by one type of cell. We were dealing with cells that differed in size with basophilic, cloudy plasma and slightly broken up nuclei with very large nucleoli, and extensive polyploidy. On the other hand, normal medullary parenchyma can hardly be found. Leukopoiesis is at times anaplastic and erythropoiesis is megaloblastic to some extent. Thrombopoiesis is not detectable. Clinical assessment: myeloma (Dr. Goll).

Röntgenologically there is no basis for this support of the skeletal system. The local findings do not improve with penicillin therapy.

Antrotomy left: Typical retroauricular incision. After the soft portions and the periosteum is pushed aside the mastoid process is exposed. It is opened by means of a hammer and chisel. A greenish serous secretion pours out freely. The enlarged cell system with the septum completely intact is reamed out. The large cells at the tip of the mastoid are especially profusely filled with the above mentioned greenish secretion. The atrium is opened wide. Marbada powder is introduced, tamponade, and the wound is closed to two-thirds.

Summary: We are dealing here with an otitis media with the clinical symptoms of mastoiditis. In the antrotomy we did not find any free pus, but rather a greenish serous secretion and a clearly thickened mucus membrane which we subjected to histological examination.

The secretion was examined cytologically and the following findings were made: the smear contained many individual and clumped cells of various sizes and slightly broken up for the most part oval nuclei some of which contained nucleoli and some of which were polymorphic. The plasma is bright basophilic and vacuolic. In addition, there are isolated lymphocytic cells, granulocytes could not be found. Macrophages were very rare.

Clinical assessment: No basis for inflammation. In the cells that were described we are dealing with myeloma cells which, of course, vary morphologically from the myeloma cells found in the bone marrow (Dr. Goll).

The histological examination of the postoperative material resulted in the following: the bone is inconspicuous. The mucous membrane sample clearly shows that it is not inflamed. Infiltrations of individual or small groups of very large cells which are greatly changed due to having been crushed and they cannot be classified with certainty. From the comparison with the sternal puncture and smears from the operating site which were stained according to Giemsa (Dr. Goll) we found that we are dealing entirely with immature plasma cells enlarged nucleolus as in bone marrow.

Diagnosis: Myelomatous infiltration of the mucous membrane of the middle ear (Prof. Dr. Bahrmann).

Following the normal postoperative period the patient was placed in the medical clinic.

Laboratory findings: Hemoglobin 8.6 g/100 ml, erythrocytes 2.64 million, hemoglobin_E 33 WE, leukocytes 3300; differential blood picture: basophile 0, eosinophile 2, immature 0, stab-like 17, segment-like 26, lymphocytes 48, monocytes 6, thrombocytes 81,530. Drop in blood pressure 70/106, total protein 9.8 g/100 ml. Electroprophesis: albumin 38.7%, α_1 5.2%, α_2 5.7%, β 6.9%, γ 43.5%. Narrower γ -gradient.

In the urine there is no protein, and no Bence-Jones protein. The X-ray examination of the thorax and the gaster as well as again examining the skeleton roentgenologically did not show any plasmacytomas or neoplastic processes. Since there was no complaint of plasmacytomas, cytostatic or X-ray therapy was disregarded. Only a blood transfusion was given. In addition, the circulatory system was treated with digitalis and oxyethyloxythephyllyne. The hemoglobin increased to 9.8 g/100 ml, the erythrocytes, however, did not increase, but rather remained at 2.71 million. The displacement to the left returned to 7 stab-like. The drop finally amounted to 68/105. Patient was discharged January 10, 1962.

Readmitted March 28, 1962. Hemoglobin was 7.2 g/100 ml and erythrocytes had decreased to 2.59 million. In the blood there was a clear shift to the left. Blood pressure increased to 120/134, total protein to 11 g/100 ml, gamma globulin to 44.6%. In the urine hyaline cylinders were now found, protein was slightly positive and the Bence-Jones protein could not, however, be detected. The treatment again was blood transfusion. Thereupon the blood picture improved with 9.6 g/100 ml hemoglobin and 3.22 million erythrocytes, blood pressure dropped to 70/110. On April 12, 1962 the patient was again dismissed.

On June 19, 1962 the patient passed away due to pneumonia at the Oskar-Ziethen Hospital.⁵

⁵We would like to thank the Chief Medical Officer Dr. Zeisewitz for releasing to us the epicrisis and the postmortem results.

Excerpts from the postmortem findings: Diffuse undeveloped plasmacytomas, gray-red bone marrow in the femoral shaft and in the vertebral bodies. Massive general osteoporosis, and massive general anemia. Localized pneumonic invasion in the inferior lobe of the left lung. In the right inferior lobe there was a partial dense confluent pneumonia.

Histological findings: Spinal marrow: diffuse infiltration of the bone marrow with relatively large cells with a foamy basophilic colored plasma which contained a large nucleolus in their vesicular nucleus and showed a certain similarity to myeloblast (immature cellular plasmacytomas).

Epicrisis

We are dealing here with a diffuse plasmacytoma with an extramedullary mucous membrane plasmacytoma of the mucous membrane in the middle ear. Roentgenologically there was no real support for skeletal involvement. The protein content of the blood increased up to 11 g/100 ml in connection with which there was a typical shift, steep gradient in the area of the γ -fraction with a relative percent of 44.6. The diagnosis was made by means of sternal puncture as well as cytological and histological examinations of the postoperative specimens from the antrotomy which was performed due to mastoiditis. The blood picture showed anemia and a shift to the left which varied in strength without leukocytosis. The patient died from pneumonia the diagnosis was verified by means of an autopsy.

Discussion

The first report on mucosal plasmacytoma was given by Schridde (44) in the year 1905 in which he described a plasmacytoma in the nose and throat region. In 1942 Jäger (quoted by Bargan and Weber [41]) represented the opinion that these mucosal tumors appear very seldom and according to his examinations only 0.8% of all the tumors were of the upper respiratory and alimentary track. In the meantime,

however, reports on this subject had become so frequent that a complete review is no longer possible. Mucosal plasmocytomas are described of the nasal cavity, the nasopharynx, the larynx and the trachea, the mucous membrane of the oral cavity, the gums, the paranasal sinuses and the tonsils (1-17, 19, 20, 23-27, 30, 32, 34-40, 45, 47-50, 52). Plasmocytomas are also found among tumors of the orbita (22, 31, 42). The general opinion exists that these extramedullary mucosal plasmocytomas are relatively benign and over a period of years, even as long as a decade, they show a progressive course (25, 36). They are supposed to respond well to X-ray therapy. Treatment for several years is not unusual. Most of the cases were described without the routine clinical symptoms of diffuse plasmocytomas, such as bone marrow changes, γ -globulin production, urinary findings, especially the Bence-Jones albuminoid and osteolytic changes in the skeleton. The relationship to diffuse plasmocytoma is, therefore, not uniformly determined. Unfortunately we must differentiate two forms of plasmocytomas in the nose, throat and ear regions one such plasmocytoma which, within the framework of a diffuse myeloma, metastasizes in the facial bones and filters out of the mucous membranes in the bones (16, 22, 28, 29, 31). Here, as a rule, the more or less strongly marked clinical symptoms of diffuse plasmocytomas occur. On the other hand, there are the solitary extramedullary mucosal plasmocytomas with a more favorable prognosis. Hargon and Weber (4) discussed quite an interesting pathogenesis for these tumors. Proceeding from the generally not unquestioned concept that the plasma cells are the site for the syntheses of normal and pathological proteins and antibodies, in comparison to other mucosal areas they indicate in a normal manner the conspicuous reproduction of plasma cells in the mucous membranes of the nose-throat area and in the respiratory track from the steady contact with antigens which are carried to the area of the mucous membrane with the respiratory air. This plasma cell reproduction according to the adaptive hyperplasia by Bünzger should then finally lead, in individual cases, to benign mucous membrane plasma cell tumors. Karzov (cited by Hargon and Weber (4)) in plasma cells of the normal mucous membrane of the respiratory track already found the same cytomorphological criteria as in plasmocytoma cells. This theory could explain the frequency of the extramedullary mucosal

plasmocytomas in the upper respiratory track including the area of the nose and throat. This idea was also represented by Gastpar (20) who found 175 out of 215 plasmocytomas in the upper respiratory passages, as reported in the literature. Of course, the question remains open as to why also a large number of these cases finally end up as diffuse plasmocytoma. These two forms - the primary osseous and the mucous membrane tumors - in many cases are not clearly defined from each other. In the case described in this article, on the basis of the histological findings, a bone involvement, at least at the site of the operation, can be clearly excluded. The plasmacellular infiltration is limited to the mucous membrane. In this sense we are dealing, therefore, with an extramedullary mucosal plasmocytoma. On the other hand, all of the symptoms of a diffuse myeloma with typical findings in the bone marrow and blood protein, anemia and paraproteinemia, of course, without Bence-Jones proteinuria and without any definite osteolytic changes in the skeletal system. The course described extended over a period of three years in which anemia had already existed for five years but cannot in any way be considered as being benign. The patient was never healthy and anemia as well as paraproteinemia became progressively worse. Stobbe (46) briefly stressed the nosological entity of the solitary, multiple and diffuse plasmocytoma.

Death due to pneumonia cannot be disregarded as being the underlying cause of the illness. Martin and Johnson (33) in studying 38 causes of death due to plasmocytoma, pneumonia was present 8 times and in 9 cases pneumonia was combined with renal failures. Glöcher (21) in 51 cases of plasmocytoma found that 55% pneumonia was in the history of the patient, and 33% of the patients died due to pneumonia. Pneumonia, therefore, represents the most frequent causes of death. The differential diagnosis of an infection, in this case otitis media, is not singular. Chase (8) described a plasmocytoma with symptoms of sinusitis frontalis, Parks (37) sinusitis maxillaris and Reddie (40) a fibroid tumor. In the case reported by Remaggi and Galetti (41) in the causality, reference to a diffuse plasmocytoma was not present, of course, the authors limited themselves to describing the local findings. Histologically, the authors found the same picture as in

the case described here. The course was obviously malignant, the patient was examined the last time six years after the antrotomy with the exception of finding a slight auditory impairment pathology was not detected.

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