Medical Memoranda

Presumed Plasmacytoma of Clivus Producing Isolated Hypoglossal Nerve Palsy

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Plasmacytoma of the skull base is rare and a case associated with an isolated 12th cranial nerve palsy is presented here.

CASE HISTORY

A 44-year-old housewife presented in December 1966 with a two-month history of neck pain accentuated by coughing and sneezing. The neck was short and movements were painful, but there were no other abnormal signs. X-ray pictures of the cervical spine and chest were normal. The haemoglobin was 10.8 g./100 ml.; white cell count 6,000/cu. mm.; E.S.R. 44 mm. in the 1st hour; M.C.H.C. 32%; and a blood film showed some anisocytosis and microcytosis.

Two months later the pain was unaltered and wasting and weakness of the left half of the tongue were noted. She was admitted to the Wessex Neurological Centre under the care of Dr. G. S. Graveson for investigation. Dysarthria was obvious and she stated that since September 1966 friends had commented that she spoke as if "her plate was loose." E.N.T. examination excluded clinical 9th or 10th nerve involvement. Skull x-ray films showed the basisphenoid and body of the sphenoid to be extensively eroded, with early basilar invagination. Tomographic cuts of the midline structures failed to show any part of the clivus intact. The anterior end, floor, and dorsum of the pituitary fossa were normal, but the bone was completely missing from the level of the posterior clinoid processes to the anterior rim of the foramen magnum. The basilar impression was presumed secondary to the clivus erosion. Air encephalography and tomography were normal except for the pontine cistern, which showed an abnormal convex configuration to the upper part of the clivus.

Chest x-ray examination was negative. Haemoglobin 12 g./100 ml.; white cell count 6,300/cu. mm.; E.S.R. 132 mm. in the 1st hour; and albumin 2.5 g. and globulin 5.6 g./100 ml. Electrophoresis showed reduced albumin and alpha- and beta-globulins, with a compact band in gammaglobulin. Sia's test was negative. Sternal marrow produced a dilute specimen with plasma cells comprising 5% of all nucleated cells. Urine protein was less than 10 mg./100 ml. and Bence Jones proteinuria was not detected. Reiter's protein complement fixation test was twice and the Wassermann reaction three times anti-complementary, and Price's precipitin reaction was negative three times. C.S.F. contained 16 lymphocytes/cu. mm.; protein 35 mg./100 ml.; no excess globulin; and glucose 48 mg./100 ml. Serum calcium was 8.8 mg./100 ml.; phosphate 2.7 mg./100 ml.; alkaline phosphatase 10 K.A. units; fasting glucose 76 mg./100 ml.; and cortisol (9.35 a.m.) 9 µg./100 ml. Total urinary neutral 17-oxosteroids 4.2 mg./24 hours, total urinary neutral 17-oxogenic steroids 8.6 mg./24 hours, and total urinary free cortisol 765 µg./24 hours.

A diagnosis at that time was made of a solitary plasmacytoma destroying the skull base and directly involving the left hypoglossal nerve. She was given 4,500 r tumour dose of radiation to the skull base. Pain was reduced but the physical signs were unaltered.

In June 1967 she was admitted to St. Mary's Hospital, Portsmouth, with a one-week history of fever and vomiting. She was drowsy, confused, disorientated, and incontinent. There was neck stiffness and Kernig's sign was positive. Lumbar puncture produced cloudy fluid at 140 mm. pressure containing 1,200 white cells/cu. mm. (95% polymorphs, 5% lymphocytes); protein 300 mg./100 ml.; and sugar 66 mg./100 ml. but no organisms or acid-fast bacilli. Haemoglobin was 80%, white cell count 9,800/cu. mm., and E.S.R. 53 mm. in the 1st hour. A diagnosis was made of "chemical meningitis," possibly due to tumour irritation of the meninges, and treatment was instituted with sulpha-

diazine, chloramphenicol, and penicillin. She made a good recovery and was discharged 11 days after admission.

In June 1968 she was readmitted with a four-day history of headache, vomiting, fever, confusion, and incontinence. She had photophobia and neck stiffness, and the middle lobe of the right lung was collapsed and consolidated. Lumbar puncture produced fluid at 260 mm. pressure containing pus cells and Gram-positive cocci, and later pneumococci were grown. She was treated with penicillin and sulphadiazine.

Haemoglobin was 70%; white cell count 7,100/cu. mm. (neutrophils 4,828, lymphocytes 2,130, and monocytes 142/cu. mm.); E.S.R. 12 mm. in the 1st hour; albumin 3.38 g./100 ml.; alpha-1 globulin 1.25 g./100 ml.; alpha-2 globulin 2.00 g./100 ml.; beta-globulin 1.25 g./100 ml.; gammaglobulin 4.62 g./100 ml.; calcium 8.9 mg./100 ml.; phosphate 2.0 mg./100 ml.; sodium 124 mEq/l.; potassium 3.7 mEq/l.; chloride 99 mEq/l.; bicarbonate 22 mEq/l.; and urea 24 mg./100 ml.; serum aspartate aminotransferase was 20 and serum alanine aminotransferase 24 units; bilirubin 0.3 mg./100 ml.; alkaline phosphatase 4 K.A. units; thymol turbidity 0; and zinc turbidity 0. Immunoelectrophoresis showed the M protein to be of IgG specificity with some IgA present, but IgM was not detected.

Sternal marrow was highly cellular. Granulopoiesis showed greatly increased activity at all stages of maturation and was normal in morphology. The myeloid-erythroid ratio was about 8:1. Erythropoiesis was normoblastic in maturation and morphology. About 6% of marrow cells were atypical pleomorphic plasma cells 15-30 µ diameter. Several giant multinuclear forms and an occasional plasmacytoblast were seen. Megakaryocytes were present in apparently normal numbers. Urine contained no Bence Jones protein.

Ten days after admission repeat lumbar puncture produced fluid containing 15 white cells/cu. mm., 25 red cells/cu. mm., protein 45 mg./100 ml., and no organisms. Two days later when bending she developed severe central back pain of sudden onset without radiation. The only additional physical sign was tenderness in the dorsolumbar spine. X-ray examination showed deposits, presumably myeloma, in D 12 and possibly D 8, and further pictures eight days later showed collapse of D 12 and L 2. She was given cyclophosphamide 100 mg./day, but this was soon discontinued owing to leucopenia, and stilboestrol 20 mg./day was substituted. She continued to take this after leaving hospital. In January 1969 melphalan 2 mg./day was added but discontinued after one month owing to leucopenia. The neurological signs remained unaltered throughout.

COMMENT

This patient's plasmacytoma was associated with sternal marrow and peripheral blood changes of generalized disease at the time of first diagnosis and she later developed spinal deposits. The particular interest of this case lies in the unusual site of origin and the isolated cranial nerve palsy produced. Clarke (1954) reviewed the literature and found 24 cases of plasmacytoma of the skull base producing cranial nerve palsies and added a case of his own. Of these 25 cases three had 12th cranial nerve palsy, but each had other cranial nerve involvement also.

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REFERENCE

Clarke, E. (1954). *Brain*, 77, 61.