

ROSAI-DORFMAN DISEASE WITH INFRA-TENTORIAL LOCALIZATION: A CASE REPORT

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SUMMARY

Rosai-Dorfman disease (RDD), or sinus histiocytosis with massive cervical lymphadenopathy, occurs mostly in children and young adults. RDD extremely rarely affects the infratentorial region, without involvement of other anatomical structures. We report a case of a 36-year old woman with isolated RDD in the cerebellum. Clinical features were presented with a neocerebellar syndrome. Brain MRI showed a lesion with varying density, located in the left cerebellar hemisphere. No extracranial lesions were detected. The patient was operated on, and recovery was full. Microscopically, the process presented with lymphoplasmatic and multinucleate histiocytic infiltration. The prognosis is good.

Key words: Rosai-Dorfman disease. cerebellar tumor, surgery

Rosai-Dorfman disease (RDD), also known as sinus histiocytosis with massive cervical lymphadenopathy is a very rare condition (probably less than 1000 cases reported in the literature) of unknown etiology. It commonly presents as massive, painless, bilateral lymph node enlargement in the neck with fever. Most cases occur in the first or second decade of life and have a predilection for blacks. It was first described by Rosai and Dorfman in 1969. They reported 4 cases of a disorder diagnosed as „malignant reticuloendotheliosis”. The disease can involve lymph nodes (nodular form) or the respiratory system, nasal sinuses, eye-orbits, bones skin (extranodular form). Nodular cases have been reported, with extranodular localizations. Intracranial localization of the disease without involvement of other sites is a very rare. Clinical presentation in 90% of the cases is lymphadenopathy, but there are cases where it can be asymptomatic, painful, anemia (normo- or hypochromic microcytosis) with antibodies against erythrocytes (polyclonal hypergamaglobulinemia) was registered in 90% of the cases. Diagnosis is based on histological investigations. Lymphoplasmatic cells are differentiated, though not atypical in shape. The cytoplasm of some histiocytes is pale and eosinophilic, or yeasty. Electron microscopy reveals phyllopods protruding from membranes of all histiocytes.

The abovementioned facts prove that diagnosis based on clinical features alone is impossible. Pathomorphologic differential diagnosis includes lymphoma, plasmatic cell granuloma and Langerhans cell histiocytosis. The most difficult case for differentiation is plasmatic cell granuloma without lymphocyte phagocytosis. In the latter case, the diagnosis is based on immunohistochemistry: large pale histiocytes are positive to S-100 protein and negative to CD1a, similarly to Langerhans histiocytosis.

The prognosis is good, with long remissions. Rarely, the disease takes a severe course with a lethal outcome- in a series of 43 patients, only three (4.7%) died (1,3,5,6,8). It is noteworthy that kidneys, liver and the respiratory system were severely damaged in these patients. In most cases, treatment is not necessary because of benign development of the disease. However, if a tumor mass is detected, surgery is the treatment of choice, and in the cases of nodular form of the disease, radiotherapy or chemotherapy are administered.

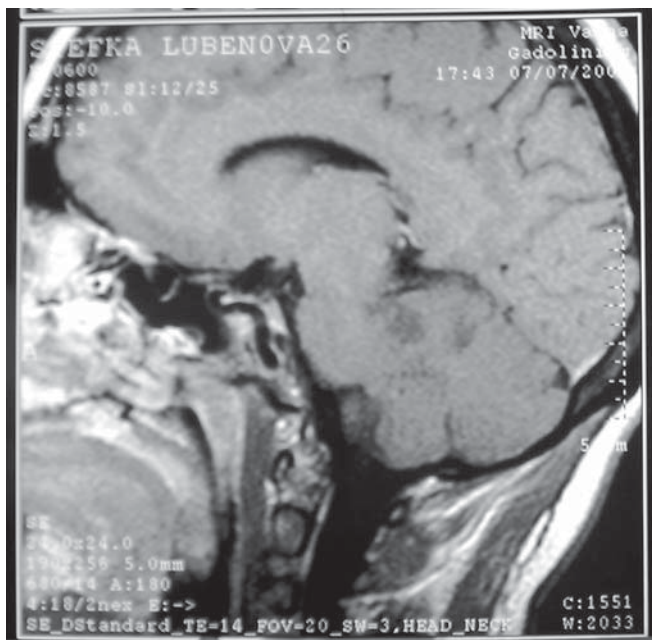
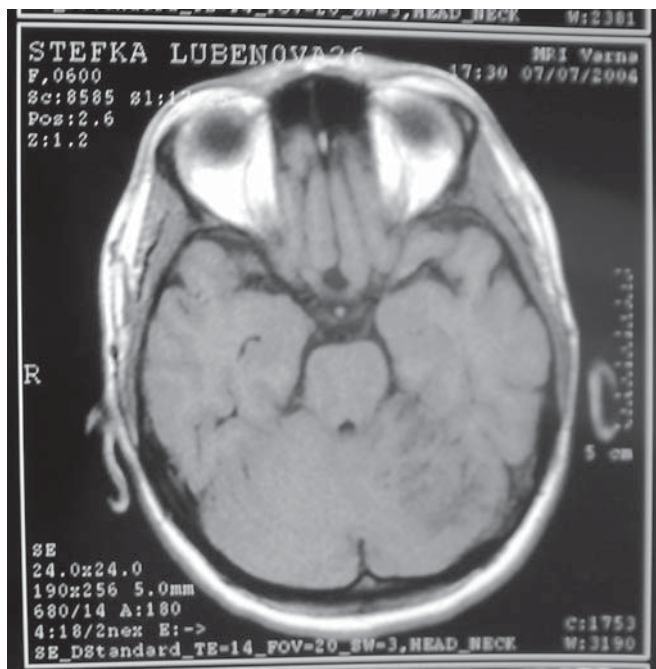
We report a 36-year old female patient with intracranial sinus histiocytosis, who reported the onset of unusual occipital headaches, nausea, vomiting, tinnitus, vertigo and seizures without losing consciousness. On examination, meningoradicular and neocerebellar syndrome were found. CT scan and MRI revealed three low-density areas with mild perifocal oedema at the fourth ventricle and the mesencephalic cistern. Introduction of contrast material did not enhance changes in images obtained. The structure visualized was suspected as a glial cerebellar tumor, and surgical treatment was agreed on.

Left-side suboccipital craniectomy was performed. At the upper lateral quadrant of left cerebellar hemisphere at approximately 5cm in depth a tumor was found. The tissue was grey in colour, poorly vascularized, with soft consistence and containing small cystic structures. The tissue was totally excised. Histopathological investigation (ref. №5322,23/16.7.2004) proved polynuclear histiocytic proliferation. Recovery was uneventful, and the neurologic symptoms disappeared. No neurological symptoms were registered at two-year follow-up examinations. However, control CT-scan showed a small low-density zone at the site of the surgical intervention.

CONCLUSION

Intracranial localizations of extranodal form of RDD are very rare (1-6, 8, 9). Most often, infiltrations in such cases are found in the dura mater, with clinical presentation mimicking meningiomas. Unlike meningiomas, infiltrations are visualized as low-density lesions. The clinical picture depends on the localization and is presented most frequently with headache, seizures and paralyzes of cranial

nerves. Despite of the fact that RDD disease is very rare, it is worth including in the differential diagnosis of meningiomas and other intracranial lesions such as glial tumors. The diagnosis based on neuroimaging methods has to be supported pathomorphologically and immunohistochemically. The treatment of choice is surgery and the prognosis is good.



Hypodense zone in left cerebellar hemispheres

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