A child with primary gastric lymphoma and cavernous sinus involvement

Primer mide lenfoması olan bir çocukta kavernöz sinüs tutulumuu

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To the Editor,

Primary gastric lymphomas are exceptionally rare gastrointestinal cancers in children [1,2]. Central nervous system involvement at the time of diagnosis is not uncommon, but there are no published reports of primary gastric lymphoma involving cavernous sinus (CS) during treatment [3,4]. We report a case of primary gastric lymphoma in a previously healthy 13-year-old boy that developed CS involvement after the second cycle of chemotherapy. He presented with a 1-month history of epigastric pain, and several episodes of hematemesis and melena. He lost 3 kg during the previous month. At presentation he looked ill, with pale conjunctiva. Physical examination showed a palpable mass in the left upper abdominal quadrant. Abdominal tomography showed diffuse thickening of the gastric rugae involving the entire fundus, a 33-mm diameter mass over the splenic vein in the corpus, and pancreatic invasion. Upper gastrointestinal endoscopy was performed and a round ulcer 3 cm in diameter was noted in the body of the stomach. The mass was diagnosed as diffuse large B-cell NHL (CD20 positive) associated with Helicobacter pylori gastritis, based on examination of the biopsy specimen. Neoplastic cells were strongly positive for CD20 and negative for cytokeratin, desmin, CD79a, S100, and CD99. The Ki67 proliferative index was 99%. EBV serology was negative. Written informed consent was obtained from the patient.

The patient was treated for the H. pylori infection using a proton pump inhibitor combined with clarithromycin and amoxicillin. Computed tomography of the chest, cranial MRI, and bone scintigraphy were normal. Bone marrow aspiration showed that there wasn't any malignant cell infiltration. He received 2 cycles of a chemotherapeutic regimen that included rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisolone. The initial cytologic status of CSF was normal; therefore, he did not initially receive intrathecal treatment.

After the second chemotherapy cycle he presented with a 2-week history of headache and left

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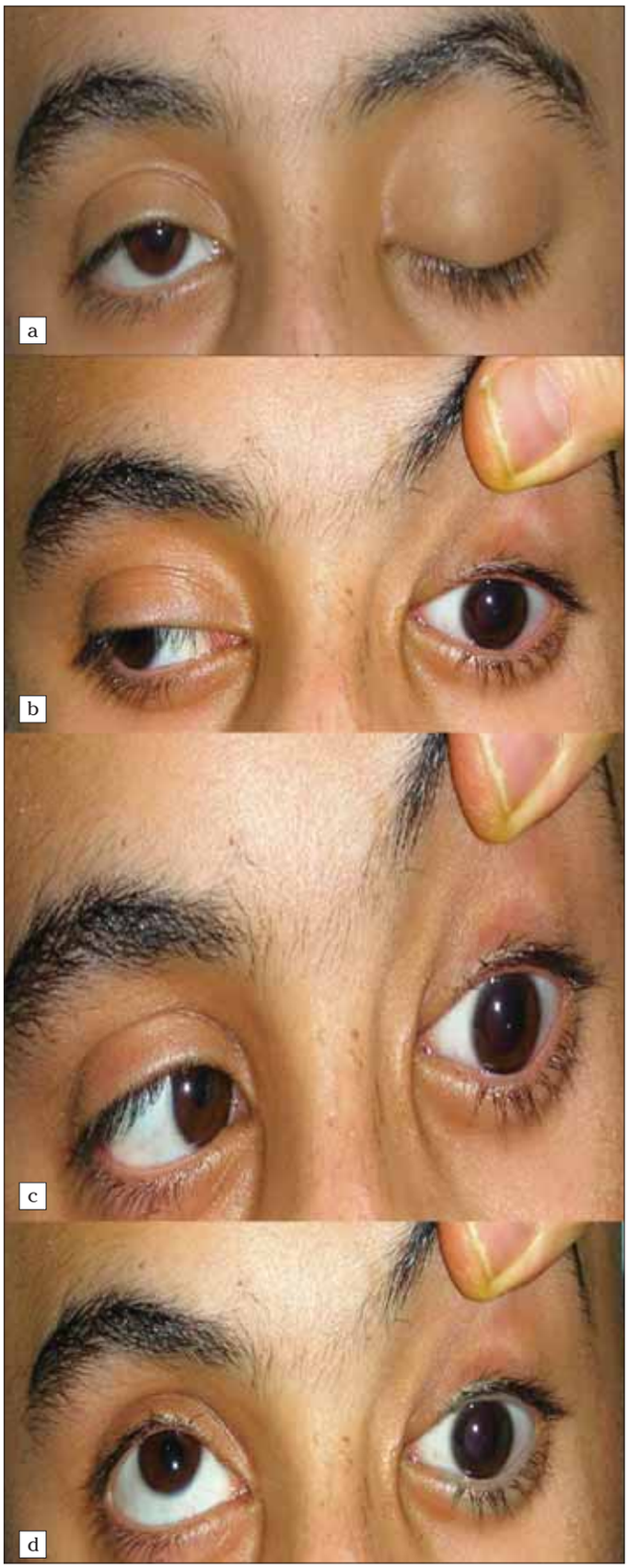

Figure 1. Left eye ptosis (a) and limitation in adduction, abduction and upward gaze of left eye (b-d)

eyeball pain. Visual acuity was 20/20 in both eyes. There was 2 mm of proptosis and complete ptosis in the left eye. Left extraocular movement was markedly limited in all directions of gaze (Figure 1a-d). MRI of the brain showed a homogeneously enhancing mass in the left CS extending into the orbital apex (Figure 2). CS syndrome was considered, based on clinical manifestations and physical findings. Lumbar puncture was performed, and cytologic examination of CSF showed large lymphoma cells with CD20 (+). Treatment was initiated with high-dose iv and intrathecal chemotherapy. Progressive ptosis of the left eye, swelling of the left orbital region with diplopia, and anesthesia developed after 3 cycles of chemotherapy. Radiation therapy was planned, but was not performed, as the patient unfortunately died 2 months after initial presentation due to progressive disease.

Primary lymphoma of the stomach is extremely rare in childhood, and is frequently designated as mucosa-associated lymphoid tissue (MALT) lymphoma in adults [5]. There is no consensus concerning the best management strategy. Surgery, H. pylori eradication, chemotherapy, radiotherapy, and combined methods have all been used for treatment. Despite the rarity of primary gastric lymphoma in childhood, it should be approached aggressively [5,6]. CS involvement with neuro-ophthalmological symptoms is even more rare; only a few case reports exist in the literature [7]. The etiology of CS syndrome is neoplasm, aneurysm, thrombosis,

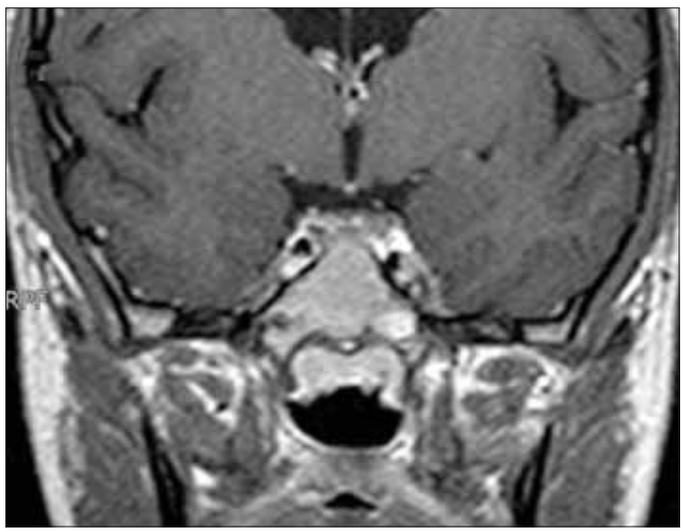

Figure 2. MRI scans were revealing cavernous sinus left side enlargement and tumor extension

carotid-cavernous fistula, pituitary apoplexy, granulomatous inflammation, and infection [4,8]. CS syndrome is suspected when retro-orbital pain is accompanied by involvement of 1 or more cranial nerves to the ocular muscles. Lymphoma may also appear as diffuse enlargement and enhancement of the CS that is similar to the appearance of metastasis. If imaging findings are not conclusive, biopsy may be necessary. A search of the literature showed that there are 13 reported cases of systemic malignant lymphoma with CS involvement, 3 of which are children [8]. To the best of our knowledge this is the first case of primary gastric lymphoma presenting with CS involvement, without an intracerebral mass or nodular lesion.

Acknowledgements

This study was performed in Erciyes University. Our case approved by the ethical committee of the institution for publishing.

Conflict of interest statement

The authors of this paper have no conflicts of interest, including specific financial interests, relationships, and/or affiliations relevant to the subject matter or materials included.

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