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Extranodal Non-Hodgkin Lymphoma Involving the Muscles of Mastication

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Summary: Involvement of the skeletal muscle by non-Hodgkin lymphoma (NHL) is unusual. The most commonly affected muscles are those of the extremities, pelvis, and glutal regions; rarely are the muscles in the head and neck involved. We report the clinical, CT, and MR imaging findings in two patients with extranodal NHL in the head and neck region involving the muscles of mastication. One patient was immunocompetent and had lymphoma that arose within the muscles; the other was a patient with AIDS who had disseminated disease at diagnosis. In both patients, the involved muscles were isodense with normal muscles on CT scans. On MR images, the infiltrated muscles were isointense with normal muscles on the T1-weighted sequence and hyperintense on the fast spin-echo T2-weighted sequence, with variable enhancement after administration of intravenous contrast material.

Extranodal manifestations of lymphoma are well recognized and occur in 20% to 30% of patients (1). Skeletal muscle involvement is uncommon. The most often affected are the gluteal and pelvic muscles, occurring as a result of metastatic hematogenous or lymphatic spread, or contiguous spread from adjacent involved lymph nodes or bone; involvement from primary disease is least common (2). Only three cases of extranodal non-Hodgkin lymphoma (NHL) that appeared to arise from the muscles of mastication in the non-AIDS patient have been reported (3–5).

We present the clinical, CT, and MR imaging findings in two cases of biopsy-proved extranodal NHL in the head and neck region involving predominantly the muscles of mastication. One patient was immunocompetent when diagnosed and the other had AIDS with disseminated disease.

Case Reports

Case 1

A 52-year-old woman presented with a painless, slowly growing mass in the right cheek, which she first noticed a year earlier. She was otherwise healthy, with no significant medical history. Clinical examination revealed a painless fullness in the right cheek region. Intraoral examination showed a right submucosal tonsillar and buccal mass that extended to the level of the third molar tooth on the right. There was no palpable lymphadenopathy.

A noncontrast CT scan of the neck showed diffuse infiltration of the masseter and medial pterygoid muscles in the right masticator space by a homogeneous mass, with posterior extension to involve the adjacent parotid gland (Fig 1A). The infiltrated muscles were isodense with normal musculature. There was no evidence of calcification or pathologic lymphadenopathy. The underlying mandible was intact with no blastic or destructive changes.

MR imaging was performed on a 1.5-T MR unit using an anterior neck coil. Spin-echo (SE) T1-weighted (500/90/2 [TR/TE/excitations]), fast spin-echo (FSE) T2-weighted (7000/156/2 [TR/TE/excitations]), short-inversion-time inversion recovery (STIR) (2000/40/2; inversion time, 150), and contrast-enhanced T1-weighted fat-suppressed images were obtained. A lesion centered in the right masticator space was seen infiltrating the masseter and medial pterygoid muscles, with relative sparing of the lateral pterygoid and temporalis muscles. The mass was isointense with normal muscles on the T1-weighted images, hyperintense on the FSE T2-weighted and STIR images, and showed fairly homogeneous diffuse enhancement after contrast administration. It extended inferiorly to the level of the angle of the mandible, wrapping around the ramus, abutting and displacing the submandibular gland inferiorly (Fig 1B and C). The right pharyngeal wall was displaced medially without significant airway compromise or gross evidence of infiltration of the fat in the right parapharyngeal space. Laterally, the mass extended through a widened right styломandibular tunnel. The lesion abutted on and was inseparable from the deep lobe of the parotid gland.

A histologic diagnosis of diffuse small lymphocytic, plasmacytoid, B-cell lymphoma was made after a buccal biopsy of the lesion. Staging CT scans of the chest, abdomen, and pelvis showed no evidence of disseminated disease. A follow-up CT scan showed complete regression of the tumor.

Case 2

A 52-year-old man with AIDS had painless proptosis of the right eye, diplopia, and headache. At his nursing home, he was initially thought to have inflammatory sinusitis and had been given a course of antibiotics with no symptomatic relief. A change developed in the quality of his voice, and he experienced decreased hearing bilaterally. His medical history included hypertension, psoriasis, hepatitis C infection, and renal
insufficiency. He was found to be HIV-positive 4 years earlier, when he was treated for an episode of meningitis.

On clinical examination, he was afebrile and had proptosis of the right eye and restricted movement of the right lateral rectus muscle. Diffuse soft-tissue swelling of the right side of the neck was also noted.

A CT scan of the paranasal sinuses, obtained after intravenous administration of contrast material, showed bilateral infiltration of the muscles of mastication. The abnormal soft-tissue mass extends to involve the sphenoidal sinus and the nasopharynx.

MR imaging of the sinuses and orbits was performed on a 1.5-T MR unit. SE T1-weighted (600/18/2), FSE T2-weighted (3600–5000/95/2 [TR/TE/eff/excitations]) and contrast-enhanced fat-suppressed T1-weighted images showed a mass lesion replacing the clivus and filling the sphenoidal sinus with extension into the apex and roof of the right orbit, causing proptosis. The lesion was isodense with normal muscles. Opacification of the sphenoidal sinus, mastoid, and ethmoidal air cells was seen. The osseous structures appeared intact with no gross evidence of erosion.

MR imaging of the sinuses and orbits was performed on a 1.5-T MR unit. SE T1-weighted (600/18/2), FSE T2-weighted (3600–5000/95/2 [TR/TE/eff/excitations]) and contrast-enhanced fat-suppressed T1-weighted images showed a mass lesion replacing the clivus and filling the sphenoidal sinus with extension into the apex and roof of the right orbit. The mass was isointense with normal muscles on T1-weighted images, hyperintense on T2-weighted images, and enhanced moderately in a nonhomogeneous manner after contrast administration. There was also diffuse infiltration and enlargement of the temporalis and pterygoid muscles bilaterally, with relative sparing of the masseter muscles, by a process with similar signal intensity and enhancement (Fig 2). Circumferential involvement of the nasopharynx down to the level of the oropharynx was noted. Intracranially, thickening and enhancement of the dura along the anterior walls of the middle cranial fossae were present.

A biopsy revealed B cell lymphoma, diffuse large cell type. A noncontrast CT scan of the chest showed several noncalcified subpleural nodules in the right lung apex but no evidence of mediastinal lymphadenopathy. However, peripancreatic, porta hepatitis, celiac axis, and retroperitoneal lymphadenopathy were noted on a contrast-enhanced CT scan of the abdomen. These findings were consistent with disseminated lymphoma.

The patient is currently undergoing a course of reduced-dose chemotherapy with methotrexate, bleomycin, doxorubicin, cyclophosphamide, vincristine, and dexamethasone (m-BACOD) with granulocyte-macrophage colony-stimulating factor (GM-CSF).

Discussion

Lymphomatous involvement of muscle has been reported to occur in only 1.4% of cases, with 0.3% occurring in Hodgkin disease and 1.1% in NHL (2). Primary muscle lymphoma is even less common (6–9). Travis et al (6) reported eight cases of primary muscle lymphoma out of 7000 malignant lymphomas seen over a 10-year period (0.11%). NHL is approximately 60 times more frequent in patients infected with HIV than in the general population (10). Ex-
tranodal presentation occurs in about 55% of AIDS-related NHL, with lymphomatous involvement of the muscle seen in 8.8%, of which 80% of cases develop primarily in the muscle (11), most commonly in the iliopsoas, paraspinial, pelvic, and extremity muscles.

Extranodal NHL arising from muscles of mastication in the head and neck region has been reported in only three cases. Harnsberger et al (3) described the CT findings in two patients who had extranodal NHL involving the muscles of mastication, but the disease appeared to be localized to the muscles in only one of the two cases. Set et al (4) described a 6-year-old non-AIDS patient with primary NHL of the acute lymphoblastic type involving the pterygomasseteric muscle complex unilaterally. Ceyens et al (5) also described a patient with extranodal NHL primarily involving the masseter muscle. In all these cases, only the CT findings were reported, as no MR imaging was performed.

In our case 1, the affected muscles were isodense with normal muscles on unenhanced CT scans, similar to the CT characteristics seen in the patient reported by Harnsberger et al (3). In the case described by Set et al (4), the involved muscles were hypodense relative to normal muscles, probably because their patient received intravenous contrast material whereas our patient did not.

At MR imaging, NHL involving skeletal muscle has been described as hyperintense or isointense relative to normal muscle on the T1-weighted sequence and hyperintense on the T2-weighted sequence (12–14). In both our patients, the signal intensity of the lymphomatous muscle was isointense with normal muscle on T1-weighted images and hyperintense on FSE T2-weighted images, similar to the signal characteristics of muscle NHL outside the head and neck region.

The patient described by Set et al (4), with primary muscle lymphoma (acute lymphoblastic type), responded well to chemotherapy. Our patient in case 1, with presumed primary muscle lymphoma (diffuse small lymphocytic B cell) extending to the adjacent parotid gland, received only local radiotherapy, and had an excellent response, with complete regression of the tumor.

The imaging characteristics of muscle NHL on CT and MR studies are nonspecific and can be seen in a variety of diseases. In the AIDS population, the differential diagnosis of multifocal bilateral masses involving masticator muscles includes bacterial myositis, idiopathic polymyositis, Kaposi's sarcoma, and rarely, metastases, all of which may have prolonged T2 relaxation times. It is particularly important to exclude bacterial myositis clinically as well as with tissue biopsy and culture. This entity is usually caused by *Staphylococcus aureus* and is potentially curable but can be fatal if not treated immediately (15, 16).

Pathologically, lymphoma can be confused with other small-cell malignancies, such as metastatic oat cell carcinoma, neuroblastoma, plasmacytoma, and rhabdomyosarcoma.

Conclusion

Awareness of NHL as a differential diagnosis in patients with and without AIDS, who present with unilateral or bilateral muscle enlargement in the head and neck region, allows appropriate handling of the tissue at biopsy. Special techniques, like electron microscopy and immunohistochemical cell marker studies, can then be used to differentiate these lesions, thus ensuring proper diagnosis and optimizing patient management (17–19).

References