

# Ectopic Lacrimal Gland of the Orbit

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A case of intraorbital ectopic lacrimal gland was evaluated with various radionuclides. CT and MRI with contrast enhancement showed a soft-tissue mass in the orbit. The lesion was gallium-avid, but  $^{18}\text{F}$ -FDG PET demonstrated very faint uptake. On early and delayed  $^{201}\text{Tl}$ -Cl images, mildly increased uptake and mild retention were observed. Technetium-99m-(V)-DMSA SPECT showed intense accumulation on both the early and delayed images. Technetium-99m-pertechnetate SPECT, demonstrating marked uptake in the lesion, was useful for the differential diagnosis between intraorbital malignancy and this benign lesion. These scintigraphic variations played an important role in the tissue characterization of this uncommon lesion.

**Key Words:** ectopic lacrimal gland; orbit; technetium-99m-pertechnetate; SPECT

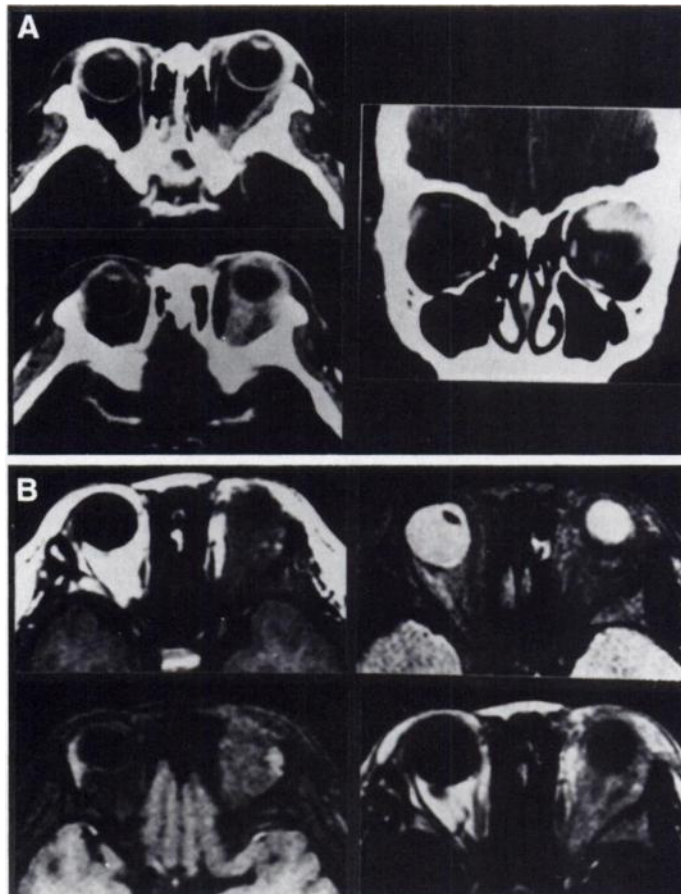
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**E**ctopic lacrimal gland, being one of the choristomas, comprises lacrimal gland tissue outside of the orbital fossa. When the lacrimal anlage is laid down by evagination of the supratemporal embryonic conjunctival epithelium into the anterior orbital soft tissues, small sequestrations apparently are able to migrate more deeply into the orbital soft tissues (1,2). This choristomatous mass causes unilateral proptosis and clinically simulate a neoplasm (3,4). Unilateral proptosis in childhood is usually an ominous symptom suggesting intraorbital malignancy, such as rhabdomyosarcoma, non-Hodgkin's lymphoma, metastatic neuroblastoma and Ewing's sarcoma. Differentiating between orbital benign lesions and malignant tumors is imperative. We describe a pediatric case of unilateral proptosis associated with intraorbital ectopic lacrimal gland, illustrating the diagnostic features of nuclear imaging.

## CASE REPORT

A nine-year-old boy had developed unilateral ocular proptosis and conjunctival swelling of 2 mo duration after an operation for left lower trichiasis. An antibiotic and a corticosteroid were administered based on a diagnosis of postoperative inflammation. The swelling was somewhat reduced but the ocular proptosis remained. On admission, the child complained of minimal discomfort in the left eye and manifested left exophthalmos. An ocular examination revealed normal visual activity but mobility of the left eyeball was slightly restricted in all directions. The right and left intraocular pressures were 20 and 29 mmHg (normal 10-21 mmHg), respectively. No abnormality was observed in the optic fundus or pupil reflex.

CT demonstrated a soft tissue mass of increasing density with contrast enhancement in the left orbit (Fig. 1A). The most likely diagnosis from the CT was intraorbital neoplasm. Axial T1-weighted (TR 500, TE 15) and T2-weighted (TR 4500, TE 100) spin-echo MR images (Fig. 1B) demonstrated an abnormal intra-

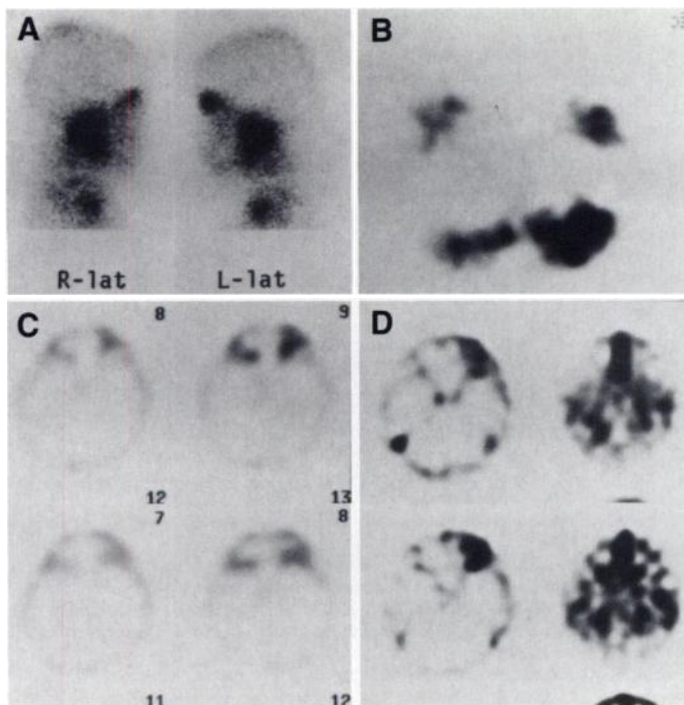


**FIGURE 1.** (A) Axial and coronal orbital CT revealed contrast-enhanced masses in the left orbit. (B) Axial T1- (upper left) and T2- weighted (upper right) MR images demonstrated an abnormal intraorbital mass with low signal intensity. On fat-suppressed T1-weighted sequences (left lower), the normal lacrimal gland is well demarcated from the mass-like lesion. T1-weighted MR images with gadolinium enhancement showed inhomogeneous enhancement of the mass (right lower).

orbital mass as an area of low signal intensity. In fat-suppressed T1-weighted sequences (TR 500, TE 20), the mass appeared to have lower intensity than the normal lacrimal gland within the lacrimal fossa. Contrast-enhanced T1-weighted MR images revealed intense inhomogeneous enhancement of the mass.

All nuclear imaging was performed with the patient's informed consent. Gallium-67-citrate scintigraphy (74 MBq intravenous, spot and whole-body images were obtained 3 days postinjection) showed an intense tracer uptake in the left superior lateral orbit, whereas faint uptake was present on the right side (Fig. 2A). Orbital PET studies with 185 MBq intravenous  $^{18}\text{F}$ -FDG were obtained at 30 min postinjection (Fig. 2B). The PET showed a very faint uptake in the left orbit. The metabolic ratio (5), which was the ratio of average counts per pixel in the left orbital mass to those in the normal cerebellum, was 0.63. Thallium-201-chloride SPECT studies were performed (74 MBq intravenous images were obtained 20 min postinjection for the early images and 4 hr postin-

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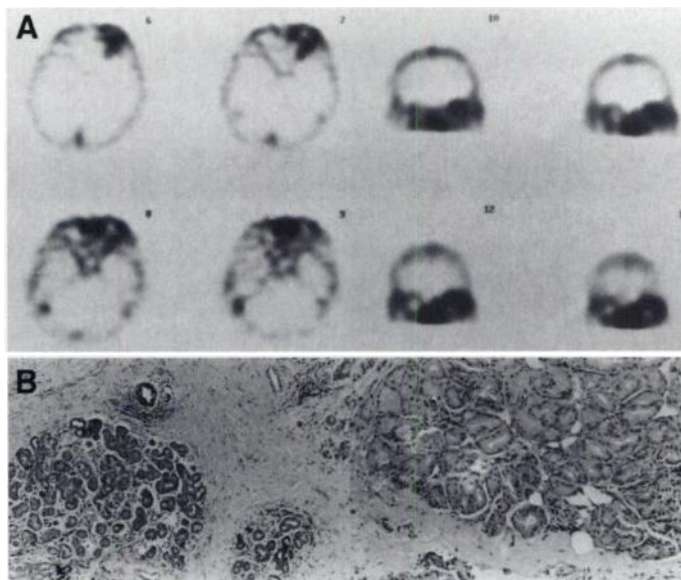
**FIGURE 2.** (A) Gallium-67 planar image showed greatly increased uptake in the left superior lateral orbit rather than in the contralateral normal lacrimal gland. (B) PET with  $^{18}\text{F}$ -FDG showed a very faint uptake in the left orbit. (C) Thallium-201-chloride SPECT in the left orbit showed slightly increased uptake on the early images (upper) and mild retention on the delayed images (lower). (D) Technetium-99m(v)-DMSA SPECT demonstrated significant uptake in the left orbit on both early (upper) and delayed (lower) images.

jection for delayed images) (Fig. 2C). Mildly increased uptake in the left orbit was seen on the early image. The early uptake ratio was 1.40 (average counts per pixel in the mass lesion to those of the contralateral orbit on the early image). Mild retention of the tracer was observed on the delayed image. The uptake ratio on the delayed image was 1.24. The retention index was 0.89 (delayed uptake divided by early uptake ratio). Technetium-99m(V)-dimer-captosuccinic acid SPECT was performed (370 MBq intravenous images were obtained 20 min postinjection for early and 4 hr postinjection for delayed images) (Fig. 2D). A significant uptake of  $^{99\text{m}}\text{Tc}$ (V)-DMSA was observed in the left orbit on both early and delayed images. The uptake ratios of early and delayed images were 2.40 and 3.25, respectively. Finally, [ $^{99\text{m}}\text{Tc}$ ]pertechnetate SPECT studies (74 MBq intravenous images were obtained at 30 min postinjection) demonstrated a highly intense uptake in the left orbital mass (Fig. 3A), representing a presence of functional glandular tissue.

Open biopsy was performed under general anesthesia to get a specimen from the orbital mass lesion, outside normal lacrimal fossa. The specimen was composed of relatively normal-looking lacrimal gland tissue with a mild inflammatory infiltrate of lymphocytes and plasma cells, histologically (Fig. 3B). The granular lobules were surrounded by thick fibrous tissue. Finally, we diagnosed this orbital lesion as an ectopic lacrimal gland on the basis of histological findings and diagnostic images. The patient showed no progression of the disease during a year without any special treatment.

## DISCUSSION

Ectopic lacrimal gland tissue is clinically uncommon. It mainly occurs in conjunctiva, limbal and cornea. Green and Zimmerman (1) reviewed 35 cases of lacrimal gland christomas, 18 of these were in the bulbar conjunctiva, six in the outer canthus, two in the lower lid, one intraocular and eight



**FIGURE 3.** (A) Technetium-99m-pertechnetate SPECT demonstrated remarkable uptake in the left orbital mass-like lesions suggesting the presence of functional glandular tissue. (B) Histopathological finding of the resected specimen demonstrated granular structure surrounded by fibrous connective tissue with some dilated ducts and with lymphocyte infiltration (left: low power, right: high power photomicroscope).

intraorbital. In our review of the literature, 22 cases of intraorbital ectopic lacrimal gland have been reported. However, there were only five articles describing the histologic picture of intraorbital ectopic lacrimal gland tissue (1,3,4,6,7). The patients with intraorbital ectopic lacrimal gland have ranged in age from 6 mo to 69 yr, but most were initially diagnosed in childhood. A more common symptom of the disease was unilateral proptosis caused by various degrees of secondary inflammation (1,3,7).

Although some reports of CT for this unusual lesion were found, there have been no reports of other diagnostic images. The differential diagnosis for intraorbital ectopic lacrimal gland is very important because this benign lesion may be incorrectly diagnosed clinically as a malignant neoplasm. With CT, it was very difficult to exclude malignancy in our patient because the CT features consisted of an irregular soft-tissue mass of increased density with various degrees of enhancement (2,4). The MR images of the lesion gave a low signal on both T1- and T2-weighted images. The low signal intensity on T2-weighted images possibly reflected the thick fibrous tissue, which was histologically confirmed within the mass in this patient.

In general, nuclear imaging has played an important role in the differential diagnosis of benign mass-like lesions from malignant tumors. We performed several nuclear imaging studies considered to have potential in tissue characterization.

Gallium-67 uptake is seen in many neoplasms. In our patient, the  $^{67}\text{Ga}$  imaging could not distinguish ectopic lacrimal gland tissue from malignancy because  $^{67}\text{Ga}$  accumulated not only in the normal lacrimal gland but also in the mass with mild inflammation that was histologically confirmed. Fluorine-18-FDG PET has been used in various malignancies, especially in brain, lung, and head and neck tumors. The glucose metabolic rates of astrocytomas, as measured with  $^{18}\text{F}$ -FDG PET, directly correlated with the histological grades (8). Recent studies revealed a relationship between glucose metabolism and proliferative activity both in vivo and in vitro (9–11). Jabour et al. (5) performed  $^{18}\text{F}$ -FDG PET for extracranial head and neck malignancies and reported that tumor metabolic ratios ranged from 0.52–1.80 (average 0.99). In this patient, the metabolic ratio

was 0.63, which was relatively low compared with the average value they reported but could not exclude a malignant tumor. Thallium-201-chloride was more specific for malignancy than  $^{67}\text{Ga}$  in a variety of tumors. Thallium-201-chloride SPECT has been used in breast and mediastinal tumors to separate benign from malignant lesions and has been used for the grading of brain tumors (12-15). The early and delayed uptake ratios and the retention index were 1.40, 1.24 and 0.89, respectively. These values probably indicated that the lesion was not a high-grade malignancy. Technetium-99m(v)-DMSA is a tumor imaging agent that classically accumulates in medullary carcinoma of the thyroid (16). More recent research has indicated that this agent can be used in head and neck (17) and lung carcinoma (18). It was reported that increased uptake of  $^{99\text{m}}\text{Tc(V)}$ -DMSA in benign lesions was very rare (17,18). However, increased accumulation of  $^{99\text{m}}\text{Tc(V)}$ -DMSA in the lesion was seen on both early and delayed images in this patient. These features seemed to contradict the histological benign findings. In our review of the literature,  $^{99\text{m}}\text{Tc(V)}$ -DMSA uptake for inflammatory masses and benign lesions has been reported (19). Technetium-99m-pertechnetate was known to accumulate in functional glandular tissue, such as in thyroid, stomach and salivary glands. Technetium-99m-pertechnetate SPECT, demonstrating high accumulations in the orbital lesion, was very useful for the final diagnosis in our patient. This finding was directly correlated with the histological specimens representing serous glandular structure with some dilated ducts, but there was a case of well-differentiated adenocarcinoma of the lung with marked uptake of  $^{99\text{m}}\text{TcO}_4^-$  (20). In addition, there were cases of ectopic lacrimal gland tissue causing two mixed tumors and an adenocarcinoma (1,21,22). These reports suggest that caution should be exercised during long-term follow-up.

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