INVESTIGATIVE NUCLEAR MEDICINE

Diagnosis of Jugular Paraganglioma by Radionuclide Angiography: Concise Communication

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Jugular paraganglioma is a highly vascular tumor, slowly growing, extending into the surrounding structures and causing otologic and/or neurologic symptoms according to its location in the jugular bulb region or the middle-ear. In our study, modified vertex and posterior head scintiangiography was used in seven cases. Scintiangiography was positive in all seven, whereas concomitant radiographic studies were limited: four of the seven gave positive findings by transmission computerized tomography (TCT). Only four patients underwent angiography, with positive results in two. Hypocycloidal tomography was positive in three cases. However, some radiographic studies, particularly TCT, may be useful in detecting local extension, bone destruction, and soft-tissue infiltration. Radionuclide angiography proved highly reliable and should be used initially whenever a jugular paraganglioma is suspected.

J Nucl Med 24: 1005–1011, 1983

Jugular paraganglioma (JP) is a tumor of the paraganglion tissue, composed of nonchromaffin cells dispersed in a highly vascular stroma with many blood vessels and sinusoids (*1*). The tumor grows either on the floor of the middle-ear cavity or beneath it, at the dome of the jugular bulb, where it may erode the tympanic floor and extend into the middle-ear. The anatomy of the region is best portrayed by a left lateral section (Fig. 1) to stress the relations of the jugular vein and middle-ear cavity and indicate the two common sites of JP tumors: (a) in tympanic cavity, and (b) in the bulb of the jugular vein (*2*).

This tumor is the second most common in the middle ear (after acoustic neurinoma) among all tumors of the temporal bone (*3*).

The JP is a benign tumor from a histopathological point of view, but it causes local destruction due to extension. Rare metatastic spread has been reported (*3,4*).

The clinical symptoms vary according to the tumor's location and extension, and may include otologic signs (hearing impairment, tinnitus, vertigo, otorrhea) and/or neurologic signs (neuralgias and cranial-nerve palsies) (*5*). Due to the tumor's slow growth and late invasiveness, early detection is difficult (*5,6*).

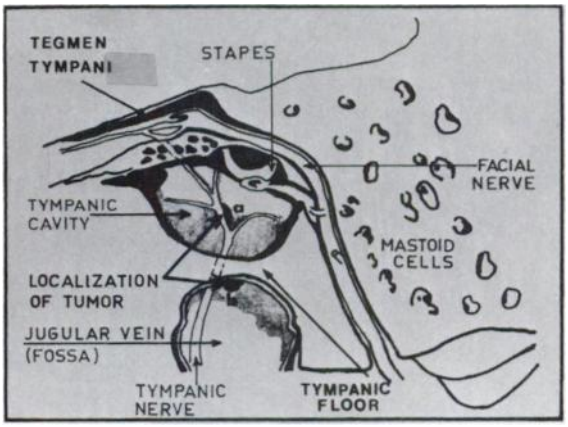

FIG. 1. Lateral section showing anatomy of region bearing jugular paraganglioma tumors. Main locations of these tumors are indicated by (a) tympanic glomus and (b) jugular bulb glomus.

Received Mar. 7, 1983; revision accepted June 20, 1983.

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TABLE 1. CLINICAL, SCINTIGRAPHIC AND RADIOLOGIC FINDINGS IN SEVEN PATIENTS WITH JUGULAR PARAGANGLIOMA (JP)

Case/ duration (years)	Age, sex	Clinical presentation Symptoms	Findings	Radiologic findings	Radionuclide angiographic findings
1. Left JP (7)	48 m	hearing loss, tinnitus, cranial palsies (VII–XII), ear bleeding	reddish mass, protruding tympanic membrane, total left-ear deafness	skull polytomography (SP): temporal partial bone destruction; computerized tomography (TCT): large hypodense mass in skull base; carotid arteriography: negative; jugular venography: jugular occlusion (Fig. 2, right)	large rounded hypervascular lesion in jugular bulb and temporal region (Fig. 2, left)
2. Right JP (2)	47 f	headaches, dizziness, hearing impairment, tinnitus	reddish pulsating mass behind tympanic membrane; sensorineural hearing impairment (30 dB)	SP and TCT*: negative	small rounded hypervascular lesion in jugular bulb region (Fig. 3)
3. Right JP (11)	69 m	hearing impairment, otorrhea, otalgia	reddish pulsating mass behind tympanic, membrane; conductive hearing loss (50 dB)	SP: clouding of right mastoid air cells; TCT: mastoid air cells clouding; carotid arteriography: negative	rounded hypervascular lesion in jugular bulb region (Fig. 4)
4. Right JP (9)	80 f	hearing impairment, ear bleeding, facial palsy	reddish-bluish mass protruding through tympanic membrane; mixed hearing impairment (55 dB)	SP: mastoid air cells, clouding and destruction; TCT: partial temporal bone destruction with soft tissue mass (Fig. 5, right)	large hypervascular lesion, lateral in jugular bulb region (Fig. 5, left)
5. Right JP	70	otalgia, hearing impairment, tinnitus, facial palsy	reddish mass behind tympanic membrane; mixed hearing impairment (45 dB)	SP, TCT* and carotid angiography: negative	rounded hypervascular lesion, medial in jugular bulb region (Fig. 6)
6. Right JP (4)	47 f	hearing impairment	bluish mass protruding through tympanic membrane; conductive hearing impairment (55 dB)	SP and TCT: negative; carotid arteriography: jugular vein occlusion	small rounded hypervascular lesion in jugular bulb region
7. Right JP (2)	53 f	hearing impairment, tinnitus, otorrhea, dizziness	reddish pulsating mass protruding through tympanic membrane; hearing impairment (30 dB)	SP: negative; TCT: jugular foramen enlargement	small rounded hypervascular lesion in jugular bulb region

* On repeated TCT 12m later: rt. temporal vascular mass.

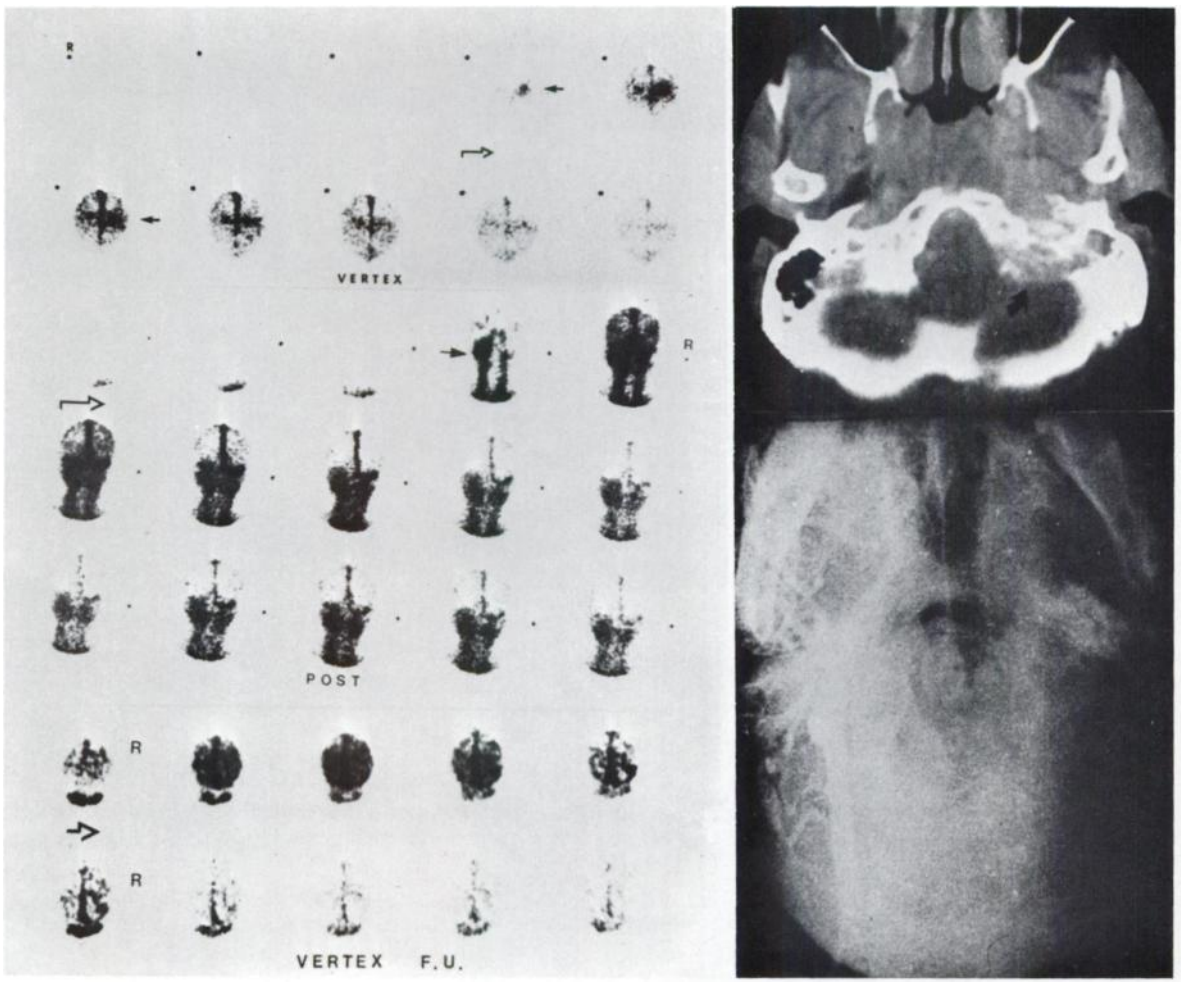

FIG. 2. Left: Jugular paraganglioma (Case 1). Vertex scintiangiography (upper) reveals left-sided rounded region of hyperperfusion in jugular-temporal region filling during arterial phase and persisting through venous phase. On posterior view (middle) same lesion is elongated in jugular bulb region. Follow-up vertex scintiangiography (lower), 2 mo after radiotherapy, demonstrates disappearance of hypervascular lesion. Right upper: TCT shows left soft-tissue mass (arrow) involving region of skull base. Right lower: Jugular venography reveals lack of filling of left jugular vein.

The advocated diagnostic imaging methods include: skull radiographs and polytomography (*7*), carotid arteriography (*8*) and/or jugularography (*9*), and recently TCT (*10*).

Anterior or posterior scintiangiography of the head has been used recently in the diagnosis of JP (*10,11*), indicating the usefulness of this radiotracer technique. However, in our experience some limitations have been encountered, particularly on anterior scintiangiography, such as: (a) unclear topographic delineation of the lesion; (b) poor distinction of JP from carotid-body tumors on arterial phase; and (c) occasional superposition of other adjacent anterior structures on the venous phase (e.g., salivary glands).

To overcome these difficulties, we used the technique of combined vertex and posterior scintiangiography. This paper describes the findings in seven cases of JP examined by this technique during the past year.

MATERIALS AND METHODS

A group of seven patients (six female, one male, aged 46–80, average 59 yr), clinically suspected of having JP, was examined. All patients underwent the following examinations: (a) otologic and audiologic tests; (b) neurologic survey, with emphasis on the cranial nerves; (c) four radiological studies (skull radiography, polytomography, CT, and angiography); (d) a radionuclide study.

The radionuclide study was performed with a rapid gamma camera equipped with a low-energy, high-resolution (10,000 parallel holes) collimator. The patients were examined in vertex and posterior positions. The aims were: (a) to demonstrate by dynamic and computerized procedures the early arterial and venous stages of the vascular flow in the jugular bulb region; and (b) to locate the vascular lesion by this two-dimensional study.

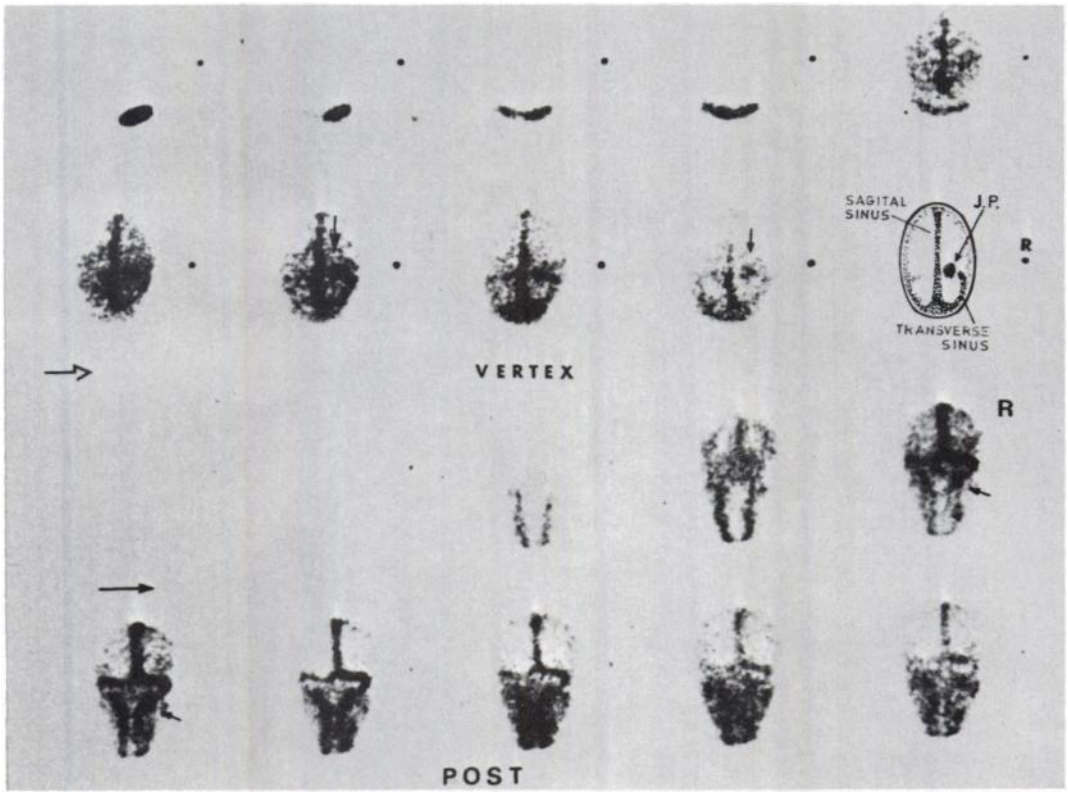

FIG. 3. Right JP (Case 2). Vertex scintiangiography shows right rounded hyperperfusion spot in jugular bulb region (arrows, upper). Vascular anatomy of JP is shown in diagram. Below: Posterior scintiangiograms show same vascular lesion in region of right jugular bulb (arrows).

The procedure started with a vertex view examination. For further details such as shape, dimension, and cephalo-caudal topography of the lesion, a second dynamic study was performed. Each study included an intravenous bolus injection of 15 mCi of pertechnetate (Tc-99m) followed immediately by dynamic imaging in a rapid sequence of 2-sec frames during the first minute after injection. The dynamic study was followed by ordinary static head scintigraphy in four views. The data of each study were fully transferred on line to the computer and stored on the disk for further image processing and clarification of the findings when necessary. All seven patients underwent these studies without any complications.

RESULTS

The personal and clinical data, the examinations performed, and the results obtained in the seven cases of JP are summarized in Table 1 and demonstrated in Figs. 2–6. In all cases there was long-standing (from 2 to 11 yr), slowly progressing, partial or complete deafness, tinnitus, otalgia, and otorrhea, with vascular masses observed on otoscopy. All were clinically suspected of

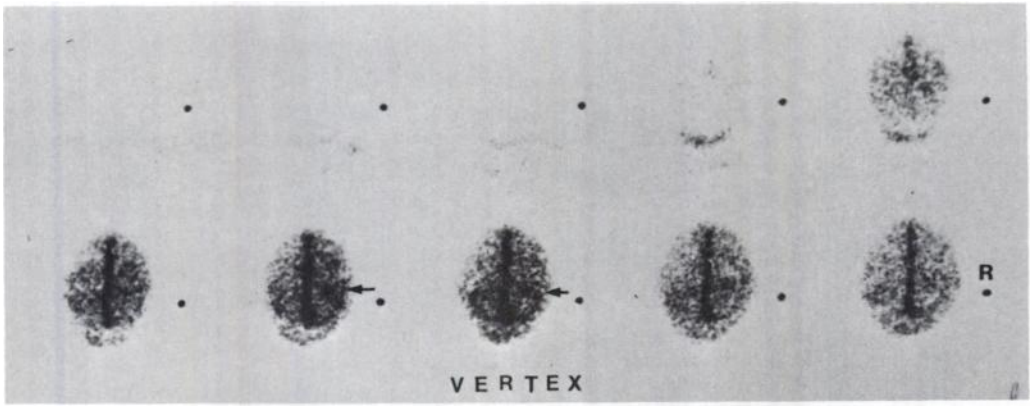

FIG. 4. Right JP (Case 3). Vertex scintiangiography shows rounded hyperperfusion spot in jugular bulb area (arrows), filling in early venous phase and clearing rapidly.

having vascular tumors in the middle ear.

To verify the diagnosis, radiographs, radionuclide, and histopathologic studies were performed.

The radiologic studies gave the following results: (a) skull radiograph and temporal tomography showed bone destruction in three of the seven cases; (b) carotid and jugular angiography demonstrated occlusion of the jugular vein in two of the four patients; (c) TCT studies, performed with and without contrast medium and with 5-mm-thick slices, detected soft-tissue or temporal-bone destruction in four out of the seven patients.

Vertex and posterior scintiangiograms were positive in all cases and were characterized by two findings:

1. On vertex view, a rounded focus of increased activity appeared in the area of the jugular bulb at the junction of the lateral sinus, resembling a pinhead at the end of a curved pin (as, for example, in Figs. 3,4). The focus showed well-defined borders on both vertex and posterior views. It appeared in the early arterial phase (on the second or third frame).

2. The regions filling duration was very short, with the activity clearing during late venous phase. Thus, the diagnosis was made by the dynamic study during the first 20–30 sec after injection of the bolus. The focus cleared soon thereafter and all the static brain scintigrams were negative.

The final diagnosis of JP was confirmed histologically. Three of the patients underwent radical mastoidectomy, with partial or complete removal of the tumor. Radiation treatment, with 6,600 rad tumor dose, was administered to three other patients, and one is awaiting surgery.

There were no false-positive or false-negative findings.

DISCUSSION

Jugular paraganglioma is found predominantly in females over age 40, and on the left side (*2,4*). In this study there were six females and one male, all over age 40. Our group differed from others: six out of seven had right-sided tumors.

This tumor originates in the perineural and perivascular adventitial tissues containing nests of large nonchromaffin cells bordered by thin-walled blood vessels in an alveolar pattern, causing shunting of the venous blood with sinusoidal dilatations (*1,12*). This seems to be the reason for the clear hypervascular area of increased radioactivity appearing in the early arterial or venous phases on dynamic scintiangiography. The lesion clears rapidly resembling the pattern similar to some arteriovenous malformations (*13,14*). However, the immediate and delayed static images were negative and noncontributory in our cases.

Vertex and posterior head angioscintigrams have the advantage that they help to establish topographic location and extension of JP, which may be: (a) superficial and lateral in the middle-ear cavity (tympanic glomus) as in Fig. 5 (Case 4), or (b) deep and medial in the dome of the jugular bulb (jugular glomus) as in Fig. 6 (Case 5). In most advanced cases, when the tumor erodes the floor of the tympanic cavity, the spot is visualized in a more extensive pattern, as in Fig. 2 (Case 1). This two-dimensional topographic localization is of utmost importance for the surgical approach or for radiotherapy, and was achieved by the method of dual vertex and posterior angioscintigraphy. In the few cases reported in the literature where a single anterior or posterior angioscintigram was performed (*10,11*), a diagnosis of a vascular lesion was made, without complete definition and extent of the lesion. In the other few chemodectoma cases examined with radionuclides (*15–17*), different dynamic and static imaging techniques and various compounds were used, mostly with less clear topographic delineation of the lesions. They included one case having three cervical chemodectomas, which were missed on perfusion study but were finally demonstrated by arteriography (*16*). In our JP series, the visualization of the tumor by the consecutive vertex and posterior head views resulted in high accuracy and better delineation of the location, size, shape, and extent of the tumor in the region of the temporal bone. This technique eliminated possible confusion with other cervical and skull-base

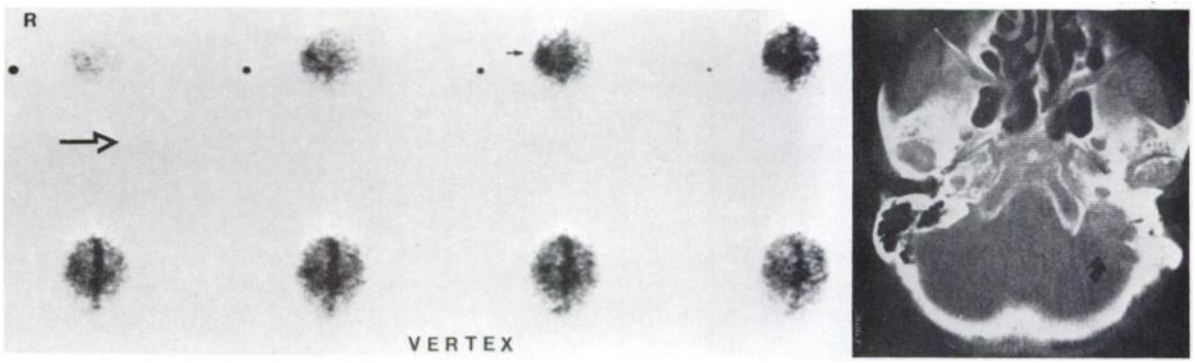

FIG. 5. Right JP (Case 4). Left: Vertex scintiangiograms show large lateral region of hyperperfusion in right jugular-temporal region (arrow), filling in arterial phase. Right: TCT shows partial destruction of right temporal bone by soft-tissue mass (arrow).

structures that may appear on anterior scintigraphy (e.g., salivary glands) as we have noted in our first cases, and was also noted by Wilson (*17*).

The differential diagnosis of JP tumors should include other lesions in the same region that are potentially diagnosable by scintigraphic studies, such as meningioma, acoustic neurinoma, hemangioma, cholesteatoma, and high carotid-body tumor. From our experience and that of others (*18*), the first three of these vascular tumors do not demonstrate the specific feature of rapid dynamic clearance of the hyperemic focus, and they do retain a focus of increased activity on static brain images. On the other hand, cholesteatomas do not show hypervascularity on dynamic scintiangiography (as in two still unpublished cases), while carotid-body tumors are anteriorly located and confined to the cervical carotid region (*19–21*). It is also important to note that a high jugular bulb may occasionally be confused with a JP tumor on otoscopy (*22*). On scintiangiography, however, the high jugular bulb differs significantly, lacking the early arterial filling and the rounded, focal, well-defined shape of the JP, as demonstrated in our posterior and vertex dynamic studies.

The growth of JP is slow, and patients with large tumors have been known to live for many years, with 71% surviving 5 yr and 29% surviving 10 yr (*3*). A case of 42-yr survival has been reported (*23*). In our group the longest interval between clinical symptoms and diagnosis was 11 yr. In advanced cases, complications occur due to a wide extension, with local infiltration causing pressure, destruction and otologic and neurologic signs. At this stage the surgical treatment may be ineffective or incomplete (*3,4*). Therefore, the importance of early diagnosis should be stressed. Using vertex and posterior cranial scintiangiography, early diagnosis was achieved.

In Cases 2 and 7 the duration of symptoms before diagnosis was 2 yr, and in Case 5, 7 yr, while all the other radiological tests remained negative. In Case 7 the only positive finding was enlargement of the jugular foramen found on TCT examination.

The setback in conventional radiological methods is due to the small size and slow growth of this tumor, with no destruction of bone in the early stages (*6*). The radiological changes will vary depending upon the site of the tumor and its direction of growth, and when the tumor is small and confined to the middle ear, radiological signs may be negative (*24*). Radiological tests demonstrating clouding of the mastoid cells, enlargement of the jugular foramen, and destruction of the temporal bone or a soft-tissue mass in the area are nonspecific (*6*), but give the location of the tumor and areas of extension, adding to the vascular-topographic information obtained by scintiangiographic methods. Accordingly, other radiographic studies are also recommended when JP is suspected in order to complete the evaluation.

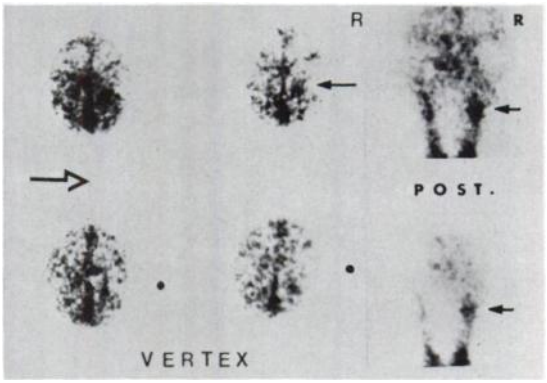

FIG. 6. Right JP (Case 5). Left: Vertex scintiangiography shows focus of hyperperfusion in medial jugular region (arrow), filling during arterial phase and clearing rapidly. Right: Posterior scintiangiography shows same jugular-bulb lesion (upper and lower arrows) in arterial phase.

A comparison of the diagnostic efficiency of the imaging methods used in our series, as well as in other JP cases reported in the literature, showed the following: skull radiography and temporal-bone polytomography are positive mainly in advanced cases (*6*), as they were in three of our seven patients. TCT may be more efficient (*10*). It was indeed positive in four of our seven cases at the time of scintiangiographic studies. Contrast angiography—including carotid arteriography and retrograde jugular venography—may demonstrate the vascularity of these tumors (*24*): in 30% of the cases by arteriography, and in 60% by retrograde jugularography (*9*). Angiographic studies were positive in two of our four reported cases (Table 1).

In conclusion, dynamic vertex and posterior cranial scintiangiography provides a highly reliable, sensitive, and accurate procedure for the diagnosis of suspected JP tumors.

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