MULTIPLE INTRAOSSEOUS HEMANGIOMAS—INVESTIGATION AND ROLE OF N-BUTYLICYANOACRYLATE IN MANAGEMENT

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Accepted 10 August 2006 Published online 17 January 2007 in Wiley InterScience (www.interscience.wiley.com). DOI: 10.1002/hed.20539

Abstract: Background. Primary intraosseous hemangiomas are rare (0.7% of all osseous neoplasms), benign, slow-growing neoplasms. These lesions are usually solitary. We are reporting a case of multicentric intraosseous hemangiomas. Investigation, treatment options, and role of N-butylcyanoacrylate (NBCA) in management will be discussed.

Methods and Results. A 20-year-old man had multicentric intraosseous hemangiomas involving the skull bones, mandible, vertebra, pelvic bone, and tibial tuberosity. N-butylcyanoacrylate was used by direct puncture technique using a transosseous transcutaneous route to control profuse bleeding from the retromolar region.

Conclusion. To the best of our knowledge, this is the first reported case with such extensive multicentric intraosseous hemangiomas. N-butylcyanoacrylate by direct puncture technique can be an effective method to devascularize and stabilize low-flow intraosseous vascular tumors.

Keywords: N-butylcyanoacrylate; multicentric intraosseous hemangiomas; direct puncture technique; Gorham syndrome; osseous hemangiomas

Primary intraosseous hemangiomas are rare (0.7% of all osseous neoplasms), benign, slow-growing neoplasms. More than 50% are found in the vertebra or skull.1 When they arise within the calvaria, they are normally confined to the frontal or parietal bones. The mandible and maxilla are the next most common sites. Life-threatening bleeding during tooth extraction or biopsy has been reported in such cases. Lesions are usually solitary and occur more frequently in females than males (ratio 3:1).2

Osseous hemangiomas are of 2 histologic types: cavernous and capillary. The cavernous type usually involves the skull.3 Only a few cases of multicentric intraosseous hemangiomas have been reported.4,5

We report a case of multicentric intraosseous hemangiomas involving skull bones, mandible, vertebra, pelvic bone, and tibial tuberosity. To the best of our knowledge, no cases with such extensive multicentric intraosseous hemangiomas have been reported in the literature. Investigation, treatment options, and the role of N-butylcyanoacrylate (NBCA) to control massive bleeding will be discussed.
FIGURE 1. Clinical photograph of the patient showing bilateral swelling in parotid region and multiple swellings involving skull bones. [Color figure can be viewed in the online issue, which is available at www.interscience.wiley.com]

FIGURE 2. CT scans of patient. (A) Expansile lytic lesion involving rami of mandible on both sides with prominent trabecular pattern. On the right side there is erosion of cortex of bone, which was site of bleeding. (B) Multiple expansile lytic lesions involving diploeic space of bone with erosion of cortex in left parietal region. (C) Polka-dot appearance is seen in upper dorsal vertebra with involvement of spinous process. (D) Expansile lytic lesion involving mandible, erosion of cortex at site of biopsy. [Color figure can be viewed in the online issue, which is available at www.interscience.wiley.com]
**CASE REPORT**

A 20-year-old man (Figure 1) was seen in the emergency department of our institute with profuse bleeding from the right retromolar region. He had a history of mild trauma to his chin. Serum calcium and phosphate levels were normal, but the serum alkaline phosphate level was slightly elevated. A CT scan (Figure 2A) showed expansile lytic lesions involving the rami of mandible on both sides with a prominent trabecular pattern. Erosion of the cortex of mandibular bone was seen, which was probably the site of bleeding. Multiple expansile lytic lesions involving diploeic spaces of skull bones were also present (Figure 2B). A polka-dot appearance was seen in the upper dorsal vertebra, with involvement of spinous process (Figure 2C). The patient had a history of biopsy of an expansile lytic lesion involving the mandible. The CT scan showed erosion of the cortex of mandible at the site of biopsy (Figure 2D).

The biopsy report revealed endothelial-lined vascular channels interspersed among fibrous bony trabeculae. The patient was sent to the digital substraction angiography (DSA) room, and NBCA (without mixing with lipiodol) was injected directly into right retromolar region. Glue cast formed at the site of the bleeding. Profuse bleeding stopped, but slight bleeding persisted from the right retromolar region.

The patient was prepared for DSA. During DSA, left external carotid artery injection showed multiple rounded areas of vascular blush in skull. Multiple small feeders from the temporal artery and middle meningeal artery supplied these areas of vascular blush. On delayed imaging, vascular blush persisted and became denser. Vividly centripetal filling of lesions along with delayed and persistent vascular blush (Figure 3) suggested low blood flow in these intraosseous vascular tumors. Right facial artery injection showed vascular blush involving the ramus of the mandible along with substracted image of glue cast injected (without contrast) prior to angiogram.

**FIGURE 3.** Digital substraction images. (A) Left external carotid injection shows multiple rounded areas of vascular blush in skull region fed by multiple small feeders from temporal artery and middle meningeal artery. (B) On delayed image, vascular blush has persisted and has become denser. (C) Image shows vividly centripetal filling of lesions. There is delayed and persistent vascular blush suggestive of these lesions being low flow vascular malformations.

**FIGURE 4.** Right facial artery injection shows vascular blush involving ramus of mandible along with substracted image of glue cast injected (without contrast) prior to angiogram.
cast due to NBCA injection (without contrast) before the angiogram (Figure 4). A microcatheter was introduced into the inferior dental artery and, for devascularization of lesion, 350 to 500 \( \mu \)m polyvinyl alcohol (PVA) particles were injected into the inferior dental artery. Bleeding from the right retromolar region persisted, so NBCA was used to devascularize low-flow vascular tumor in the ramus of the right mandible by direct puncture technique using the transosseous transcutaneous route. A vascular lesion was punctured with 22-gauge needle by transosseous transcutaneous route with strict fluoroscopic monitoring, and the correct location of the needle was verified by reflux of blood at hub. Contrast agent was injected into the lesion, revealing local parenchymography. NBCA was then mixed with lipiodol (ratio of 1:1) to make it radio-opaque. Subsequently, NBCA was slowly injected into the vascular tumor in right ramus of the mandible (Figure 5). Injection of NBCA into the intraosseous vascular tumor was monitored strictly under fluoroscopic control. For complete devascularization, multiple punctures were needed for direct injection of NBCA into the lesion (Figure 5). Bleeding from the retromolar region stopped.
The patient then underwent MRI and nuclear scans. T2-weighted MR images showed hyperintense expansile lesions involving both rami of mandible and diploeic spaces of skull bones (Figure 6). Hypointense dots showing trabecular pattern of bone were also present. Lesions were isointense on T1-weighted images (Figure 6). Tc99m (technetium 99m)-labeled red blood cell (RBC) blood pool scan showed increased radionuclide uptake at sites of bony hemangiomas. Increased radionuclide uptake suggestive of bony hemangiomas was also found in both tibial tuberosities (Figure 7). Fractionated irradiation to a total dose of 40 Gy (in 2-Gy fractions) was given for 5 weeks. The 6-month follow-up was uneventful, and the patient showed no further progression of the lesions.

DISCUSSION

Primary intraosseous hemangiomas are usually solitary. Only a few cases of multicentric intraosseous hemangiomas have been reported.4,5 Extensive and multicentric lesions involving the skull bones, mandible, vertebra, pelvis, and tibia, such as those described in this case report, have not been reported previously. Multiple intraosseous hemangiomas have been described in initial stages of Gorham syndrome (vanishing bone disease)6; however, extensive multicentric involvement such as that reported here has not been reported even in cases of Gorham syndrome (vanishing bone disease).6,7 Vanishing bone, which is characteristic of Gorham syndrome,6,7 was not found in this patient.

The CT scan demonstrated multiple expansile lytic lesions with a prominent trabecular pattern involving rami of mandible on both sides. Multiple lesions were also found involving diploeic spaces of skull bones. A polka-dot appearance was seen in the upper dorsal vertebra with involvement of spinous process. These lesions were heterogeneously isointense on T1-weighted MR images and hyperintense on T2-weighted MR images. Hypointense dots due to trabecular pattern of bone were also present.

Features of high-flow lesions (arteriovenous malformations) such as serpiginous signal voids, absent dominant mass, and decreased marrow signal on T1-weighted images8 were absent in these lesions. Therefore, on MRI, these lesions were labeled as low-flow vascular tumors. Whole-body nuclear scans detected vascular tumor in the region of iliac crests and tibia. On DSA, vividly centripetal filling of vascular tumors along with delayed and persistent vascular blushing confirmed low blood flow in these lesions. The tumors were fed by multiple small, low-flow feeders.

Low blood flow and the small size of these vessels made it impossible to negotiate a catheter through many small feeding vessels; hence these tumors (intraosseous hemangiomas) could not be devascularized completely by catheterization and...
embolization. Therefore, bleeding persisted in this case even after embolization of the inferior dental artery and conventional method of catheterization and embolization was not successful. Resnick et al9 also were not able to control life-threatening bleeding from vascular malformation of the mandible by using the traditional method of catheterization and embolization, and direct percutaneous transmandibular puncture technique of embolization was used to control bleeding.

Vascular tumor in the right ramus of the mandible in this case was also devascularized by direct puncture technique using the transosseous transcutaneous route. NBCA was used as an embolizing agent in direct puncture technique.

Because NBCA may get transported into draining vessels, this method of devascularization carries risk of anaphylactic shock10 and pulmonary embolism.11 To avoid these complications, prior to embolization, parenchymography with contrast agent should be performed, and embolization should be performed under strict fluoroscopic control. Injection of NBCA should be strictly intrallesional because superficial injection may be followed by tissue necrosis. Ethanolamine oleate, ethibloc, sodium tetradecyl sulfate, and polidoconal are other embolizing agents that can be used for embolization.12 NBCA was used as an embolizing agent in this case report because it solidifies immediately on contact with blood, so the risk of its penetration into draining vessels is minimal. NBCA has antimicrobial properties,13 tissue toxicity is minimal, and on mixing with lipiodol it forms a radio-opaque solution. If the lesion in the vertebra either grows or becomes symptomatic, percutaneous transpedicular acrylic vertebroplasty using methacrylate14 may be an option. Fractionated irradiation to a total dose of 40 Gy (in 2-Gy fractions) was applied over 5 weeks. Radiotherapy can involute or can stabilize these lesions.15 The 6-month follow-up in this patient was uneventful, without further progression of lesions.

CONCLUSIONS
To the best of our knowledge, this is the first reported case of such extensive multicentric intraosseous hemangiomas, involving skull bones, mandible, vertebra, pelvis, and tibia. 99m–labeled RBC blood pool scan can be a useful modality in screening human body for hemangiomas, and NBCA by direct puncture technique can be an effective method to devascularize and stabilize intraosseous low-flow vascular tumors.

REFERENCES