Enlarging Radiolucency of the Right Posterior Maxilla

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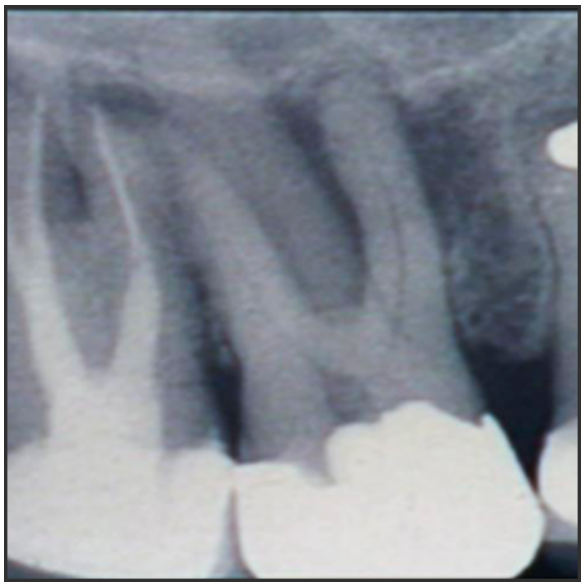
Online Case: *www.dentalcare.com/en-us/professional-education/case-challenges/case-challenge-071*



The following Case Challenge is provided in conjunction with the UT Health San Antonio School of Dentistry faculty.

An asymptomatic 49-year old male presents with an enlarging radiolucency.

After you have finished reviewing the available diagnostic information, make the diagnosis.

Diagnostic Information

History of Present Illness

Larry is 49 year-old male who presented for his 6 month dental recall examination. He was doing well and had no dental complaints. Periodontal evaluation revealed a 10 mm probing depth on the distal of tooth #2. The probing depths in this area 6 months earlier were 3 to 4 mm. A radiograph revealed a distal periradicular and apical radiolucency involving tooth #2. Tooth #2 did not respond to vitality testing and a diagnosis of a combined endodontic/periodontic lesion was established. Nonsurgical endodontic therapy and scaling and root planing were completed. A follow-up radiograph in 3 months showed no evidence of healing of the defect and the presence of a new periapical lesion around tooth #3 (Figure 1). Nonsurgical endodontic therapy was then completed on tooth #3. The patient remained asymptomatic but radiographic expansion of the lesion was noted at a three month follow-up exam (Figure 2). No cortical expansion, fistula tract, or drainage was noted. Moderate tooth mobility was present. Teeth #2 and #3 were then extracted a a fleshy tan-white tissue was curetted from the extraction sites and submitted for histologic examination.

Medical History

- Adverse drug effects: None
- Medications: fosinopril, albuterol inhaler, aspirin, fish oil, daily vitamins
- Pertinent medical history: hypertension, mild chronic obstructive pulmonary disease
- Pertinent family history: paternal - type II diabetes, hypercholesterolemia, myocardial infarction; maternal - rheumatoid arthritis
- Social history: 25 pack/year history of cigarette smoking; social alcohol use (mixed drinks); denies recreational drug exposure

Clinical Findings

The periapical radiographs of the right posterior maxilla show an enlarging destructive radiolucent lesion involving the roots of teeth #2 and #3 (Figures 1 and 2).

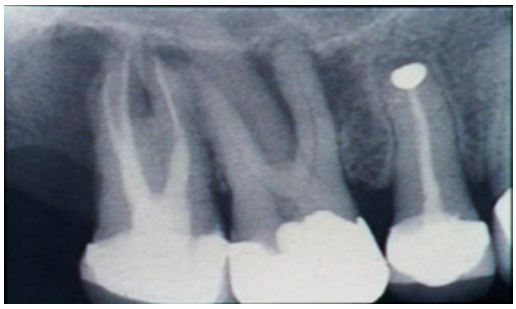


Figure 1. Three month follow-up radiograph following completion of endodontic therapy on tooth #2 showing persistence of the periradicular radiolucency associated with tooth #2 and development of a new periapical radiolucency on tooth #3.

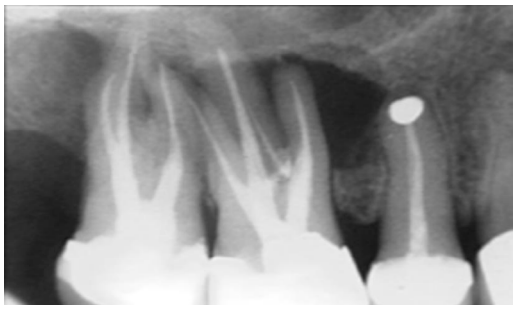


Figure 2. Three month follow-up radiograph following completion of endodontic therapy on tooth #3 showing an enlarging radiolucent lesion of the right posterior maxilla.

Histopathologic Findings

The histologic sections of the biopsy showed an infiltrate composed predominantly of histiocytes with interspersed eosinophils, lymphocytes and rare plasma cells (Figure 3). The histiocytes display bean shaped indented/ grooved nuclei and abundant pink cytoplasm. Occasional multinucleated giant cell were present (Figure 4). Immunohistochemical stains showed the histiocytes to be positive for CD1a (Figure 5) and CD207 (Figure 6).

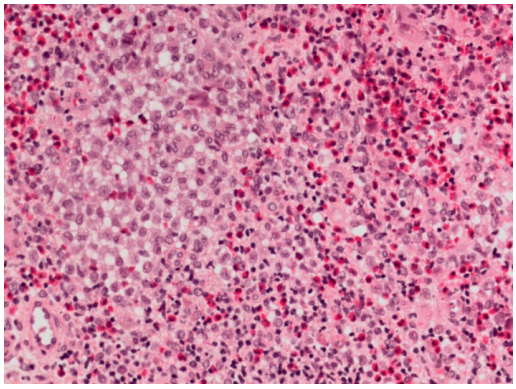


Figure 3. Low power histologic image showing a diffuse infiltrate of histiocytes with admixed eosinophils.

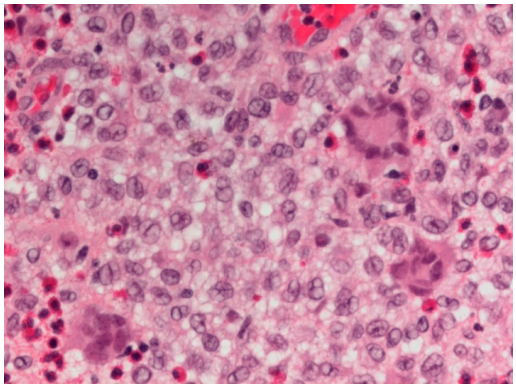


Figure 4. High power histologic image showing histiocytes with grooved vesicular nuclei and abundant pink cytoplasm. Eosinophils and focal multinucleated giant cells are present in the background.

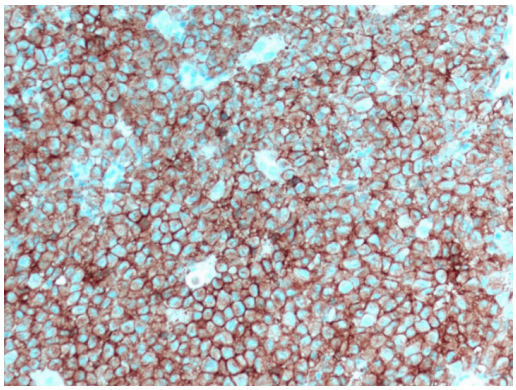


Figure 5. Immunohistochemical stain for CD1a showing positive membrane staining of the histiocytes.

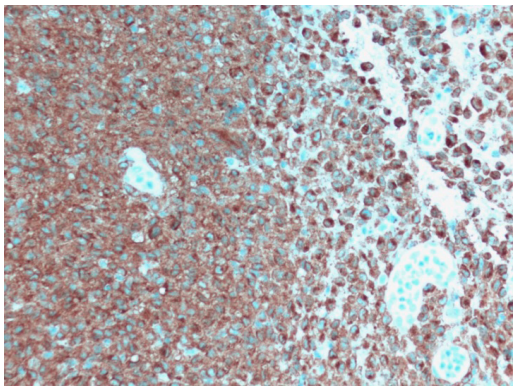
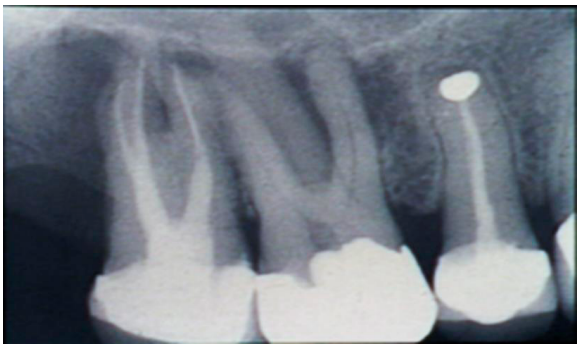


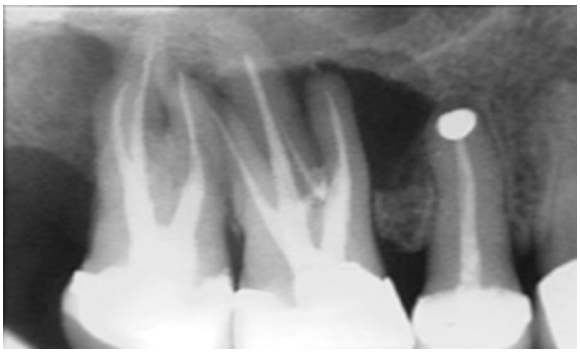
Figure 6. Immunohistochemical stain for CD207 showing positive cytoplasmic staining of the histiocytes.

Select Diagnosis

Can you make the diagnosis

A 49-year old male with an enlarging radiolucency of the right posterior maxilla.





Select the Correct Diagnosis

A. Langerhans cell histiocytosis
B. Metastatic carcinoma
C. Plasmacytoma
D. Diffuse large B-Cell Lymphoma

Langerhans cell histiocytosis

Choice A. Congratulations! You are correct.

Langerhans cell histiocytosis (LCH)[1-5] is an uncommon proliferation of dendritic macrophages that normally function as antigen processing/presenting cells in the skin, mucosa, lymph nodes and bone marrow. Many cases of LCH show mutations consistent with a neoplastic process. LCH occurs across a wide age range but is most common in children. There is not a strong gender predilection. Sites commonly involved by LCH include skin, bone, lymph nodes, lungs, liver, and spleen. LCH may show unifocal or multifocal involvement of a single organ system or disseminated multiorgan system disease with or without organ dysfunction. The skull, ribs, vertebrae and jaws are the osseous sites most frequently involved. Radiographically, the bone lesions present as a "punched-out" radiolucency. In the jaws LCH can produce a "tooth floating in air" appearance and can appear very similar to pulpoperiapical and/or periodontal disease. Soft tissue erythema, swelling and ulceration may also occur. Patients may complain of associated tenderness or dull pain. A biopsy of LCH will show a diffuse infiltrate of histiocytes with folded/grooved vesicular nuclei and abundant pink cytoplasm. Interspersed eosinophils are typically seen. Multinucleated giant cells, lymphocytes, plasma cells, foci of necrosis and hemorrhage may also be present. Langerhans cells can be identified by their histomorphology and immunohistochemical staining for S100 protein, CD1a and CD207. Electron microscopy will show characteristic intracytoplasmic rod to "tennis racket" shaped Birbeck granules. Treatment includes curettage of small accessible lesions, intralesional steroid injections, low-dose radiation therapy, and chemotherapy for disseminated disease. The prognosis is generally good for localized disease. Disseminated disease in young patients may cause significant morbidity and can result in mortality.

Metastatic carcinoma

Choice B. Sorry, this is not the correct diagnosis.

Metastatic carcinoma[1,6-8] is the most common form of cancer occurring in bone. The tumors that most frequently metastasize to bone include breast, lung, prostate, kidney, and thyroid cancers. The vertebrae, ribs, pelvis, and skull are the sites most often involved by metastatic carcinoma. Metastasis to the jaws is uncommon but may be under reported. Occasionally a bone metastasis may be the first sign that a patient has cancer. Most patients are older adults with a peak incidence in the fifth and sixth decades of life. There is no significant gender predilection. Patients may present with pain, swelling, loose teeth, paresthesia, a non-healing extraction site, and pathologic fracture. Radiographically, metastasis to bone presents as an ill-defined destructive/osteolytic radiolucent lesion. Some cancers like breast and prostate cancer may induce new bone formation and present as a mixed or opaque lesion. Tumor infiltration may produce a widened periodontal ligament space. Metastatic disease to the jaws can be mistaken for inflammatory reactive pulpoperiapical and/or periodontal disease. A biopsy will show malignant epithelial cells infiltrating the bone marrow. Special studies may be performed to determine or confirm the primary site of origin for the metastasis. Treatment may include surgery, radiation, bisphosphonates, and evolving novel biological and molecular therapies. The presence of bone metastasis indicates disseminated stage IV disease with a guarded prognosis. While the clinical and radiographic features of Langerhans cell histiocytosis may be similar, the histopathologic findings do not support the diagnosis of metastatic carcinoma.

Please re-evaluate the information about this case.

Plasmacytoma

Choice C. Sorry, this is not the correct diagnosis.

Plasmacytoma (PC)[1-2,9-11] is a solitary neoplastic proliferation of plasma cells that usually arises in bone but may also occur in extramedullary soft tissues. PC develops most often as a solitary lesion in the vertebrae of elderly males. PC may also occur in the jaws. Patients may present with complaints of pain and swelling. Radiographically PC will present as a unifocal radiolucent intramedullary lesion. Disseminated involvement of bone marrow (multiple myeloma) is not present. Extramedullary PCs commonly arise in the lungs and the head and neck region involving the nose/paranasal sinuses, tonsils, nasopharynx, or parotid gland. A biopsy will show diffuse sheets of plasma cells which may be mistaken for a reactive inflammatory infiltrate. The plasma cells produce monoclonal immunoglobulin heavy chains and one type of light chain (kappa or lambda). These immunoglobulins may be detected as a monoclonal spike on serum or urine protein electrophoresis. Immunohistochemical stains for kappa and lambda will demonstrate a monoclonal light chain restriction. These findings would support the diagnosis of a neoplastic plasma cell proliferation. Treatment for PC includes local radiation therapy and surgical resection. Unfortunately, after a number of years most patients will eventually progress to develop disseminated multiple myeloma. Extramedullary PCs have a much lower rate of progression and a better prognosis. While the clinical and radiographic features of Langerhans cell cell histiocytosis may be similar, the histopathologic findings do not support a diagnosis of plasmacytoma.

Please re-evaluate the information about this case.

Diffuse large B-Cell Lymphoma

Choice D. Sorry, this is not the correct diagnosis.

Diffuse large B-cell lymphoma (DLBCL)[1-2,12-15] is a malignant lymphoid neoplasm. It is the most common type of non-Hodgkin lymphoma in the United States. DLBCL usually arises in older adults, although it may occur in young adults and children. There is a slight male gender predilection. DLBCL presents as an enlarging mass in a lymph node or extranodal site. It represents the most common lymphoma to arise in the oral cavity. Oropharyngeal lymphoid tissue, oral mucosa (palate, gingiva, buccal vestibule) and bone of the jaws are the oral sites most frequently involved. Patients may complain of pain, swelling, loose teeth and/or paresthesia. A biopsy will show a diffuse infiltrate of large lymphocytes with vesicular round to multilobated nuclei, large nucleoli, and pale blue cytoplasm. B-cell markers CD19 and CD20 are expressed by the tumor. DLBCL is an aggressive type of lymphoma and is rapidly fatal without appropriate therapy. Treatment consists of systemic combination chemotherapy. Rituximab, an antibody against the CD20 B-cell surface antigen, has been used as adjuvant therapy. Many patients achieve complete remission and approximately 40-50% may be cured. While the clinical and radiographic features of Langerhans cell histiocytosis may be similar, the histopathologic findings do not support the diagnosis of diffuse large B-cell lymphoma.

Please re-evaluate the information about this case.

References

1. Neville BW, Damm DD, Allen CM, et al. Oral and Maxillofacial Pathology. 4th ed. St. Louis, MO. Elsevier. 2016.
2. Kumar V, Abbas AK, Aster JC. Robbins and Cotran Pathologic Basis of Disease. 9th edition, Philadelphia Elsevier/Saunders 2015.
3. Lian C, Lu Y, Shen S. Langerhans cell histiocytosis in adults: a case report and review of the literature. Oncotarget. 2016 Apr 5;7(14):18678-83. doi: 10.18632/oncotarget.7892.
4. Zinn DJ, Chakraborty R, Allen CE. Langerhans Cell Histiocytosis: Emerging Insights and Clinical Implications. Oncology (Williston Park). 2016 Feb;30(2):122-32, 139.
5. Gonçalves CF, Morais MO, de Cássia Gonçalves Alencar R, et al. Solitary Langerhans cell histiocytosis in an adult: case report and literature review. BMC Res Notes. 2016 Jan 9;9:19. doi: 10.1186/s13104-015-1799-z.
6. Pesis M, Taicher S, Greenberg G, et al. Metastasis to the jaws as a first manifestation of hepatocellular carcinoma: report of a case and analysis of 41 cases. J Craniomaxillofac Surg. 2014 Dec;42(8):1997-2001. doi: 10.1016/j.jcms.2014.09.005. Epub 2014 Sep 19.
7. Weidle UH, Birzele F, Kollmorgen G, et al. Molecular Mechanisms of Bone Metastasis. Cancer Genomics Proteomics. 2016 Jan-Feb;13(1):1-12.
8. Jehn CF, Diel IJ, Overkamp F, et al. Management of Metastatic Bone Disease Algorithms for Diagnostics and Treatment. Anticancer Res. 2016 Jun;36(6):2631-7.
9. Agostini T, Sacco R, Bertolai R, et al. Solitary plasmacytoma of the jaw. J Craniofac Surg. 2011 Nov;22(6):e2-10. doi: 10.1097/SCS.0b013e31822ec79a.
10. Canger EM, Celenk P, Alkan A, et al. Mandibular involvement of solitary plasmocytoma: a case report. Med Oral Patol Oral Cir Bucal. 2007 Jan 1;12(1):E7-9.
11. Lo Muzio L, Pannone G, Bucci P. Early clinical diagnosis of solitary plasmacytoma of the jaws: a case report with a six year follow-up. Int J Oral Maxillofac Surg. 2001 Dec;30(6):558-60.
12. Triantafillidou K, Dimitrakopoulos J, Iordanidis F, et al. Extranodal non-hodgkin lymphomas of the oral cavity and maxillofacial region: a clinical study of 58 cases and review of the literature. J Oral Maxillofac Surg. 2012 Dec;70(12):2776-85. doi: 10.1016/j.joms.2012.01.018. Epub 2012 Apr 10.
13. Djavanmardi L, Oprean N, Alantar A, et al. Malignant non-Hodgkin's lymphoma (NHL) of the jaws: a review of 16 cases. J Craniomaxillofac Surg. 2008 Oct;36(7):410-4. doi: 10.1016/j.jcms.2008.03.004. Epub 2008 Jun 17.
14. Jessri M, AbdulMajeed AA, Matias MA, et al. A case of primary diffuse large B-cell non-Hodgkin's lymphoma misdiagnosed as chronic periapical periodontitis. Aust Dent J. 2013 Jun;58(2):250-5. doi: 10.1111/adj.12056. Epub 2013 May 5.
15. Mendonça EF, Sousa TO, Estrela C. Non-Hodgkin lymphoma in the periapical region of a mandibular canine. J Endod. 2013 Jun;39(6):839-42. doi: 10.1016/j.joen.2013.01.007. Epub 2013 Mar 13.

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Since joining the faculty in 2002, Dr. Huber has been teaching both pre-doctoral and graduate dental students at the UTHSCA School of Dentistry. In 2014, he was awarded the UTHSCSA Presidential Teaching Excellence Award. He is a Past President of the American Academy of Oral Medicine. Dr. Huber has spoken before many local, state, and national professional organizations. He has published over 70 journal articles, book chapters, and online postings.

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