

June 1, 1979

CASE HISTORIES

CASE # 1. (78-15745) Contributed by Albert M. Abrams, D.D.S., M.S., Professor of Pathology, University of Southern California School of Dentistry, Los Angeles, California.

The patient, a 47-year-old chronically ill, wasted female, presented to the Dental Clinic at Los Angeles County/USC Medical Center with a 3 month history of painful enlarging mass along the right superior alveolar ridge. It was a 3 x 5 cm fungating lesion occupying most of the right upper alveolar ridge and extended onto the hard palate. Radiographs revealed no bony abnormalities. Past medical history revealed that the patient developed a soft tissue mass on the left lower extremity 4 years prior to admission. The mass was excised in Mexico, but the patient was never given the diagnosis. The leg lesion recurred and progressively enlarged until the time of admission when it was noted to be 6 x 7 cm and freely moveable. The remainder of the past medical history revealed a 3 year history of back pain attributed to an automobile accident. Anorexia and weight loss had progressed over the last 6 months. The patient had right eye pain and right sided headaches with fever during the week prior to admission. The following findings were noted on radiographic examination: (1) Multiple compression fractures of T-spine, most likely due to diffuse osteoporosis. (2) Large lobulated lesion in the left posterior lung field with erosion of posterior ribs. (3) Face, C-spine, left ankle, L-5 spine, right tibia, fibula, right and left hips were all essentially without significant abnormalities.

The submitted slide is from the oral lesion. The leg mass features essentially the same histomorphology.

CASE # 2. (04407/79) Contributed by Yvon LeGal, M.D., Institut D'Anatomie, Pathologique, Strasbourg, France.

78-year-old Caucasian female with edentulous mandible, developed a lesion in a residual cyst, radiographs are included.

CASE # 3. (S79-349 2 D) Contributed by Henry A. Azar, M.D., Chief, Laboratory Service and Professor of Pathology, University of South Florida College of Medicine, Tampa, Florida.

This hypertensive and diabetic 62-year-old male developed an infected right upper cuspid which was extracted within 72 hours. An infectious process developed around the site of extraction that necessitated close follow-up. Ten days after extraction he was transferred to this VA Medical Center where a right maxillectomy with ethmoidectomy and orbital exenteration was carried out.

CASE # 4. (79-445) Contributed by Jose M. Hori, M.D., Pathologist, Davis Memorial Hospital, Elkins, West Virginia.

76-year-old Caucasian female admitted to Davis Memorial Hospital because of trigeminal neuralgia involving the third division in the right side. The lesion was described as a swollen lower gingiva posterior to the incisor and apparently extending into or around the submandibular glands openings in the floor of the mouth. Lab and x-rays are all within normal limits.

CASE # 5. (S-79-15543, B.C.H.) Contributed by William C. Bucher, M.D., Pathologist, Boone County Hospital, Columbia, Missouri.

This 65-year-old white male first noted a nodule in the region of his left cheek 10 years previously. He was told by his family doctor that it was a "calcium deposit" and not to worry about it. Except for enlargement the patient had no difficulties until December, 1978 when his wife noted that when smiling the patient's left cheek did not move. The patient then sought medical advice and an operation was performed on 4-2-79.

CASE # 6. (S-78-2076-A) Contributed by Jonathan B. Hanson and Richard F. Graham, D.D.S., Oral Surgeons and A. Giuliani, D.O., Pathologist, Charles E. Still Hospital, Jefferson City, Missouri.

Tissue from left gingival, premolar area. This is a 5 month old infant who developed this lesion when he was 2 months old, noted by his mother during feeding. At 5 months of age the lesion was excised.

CASE # 7. (STIPE) Contributed by Charles Dunlap, D.D.S., University of Missouri School of Dentistry, Kansas City, Missouri.

This was an 84-year-old female who saw an oral surgeon because of loose teeth. X-rays showed two mandibular bicuspid teeth with partial root resorption. Molar teeth were missing. There was a 5.0 cm destructive radiolucent lesion of the body of the mandible with erosion of both lingual and buccal cortical plates of bone.

CASE # 8. (S-1219-79) Contributed by Drs. Lin and Dodhia, Truman Medical Center, Kansas City, Missouri.

This approximately 60-year-old man was seen by an oral surgeon in southwestern Missouri. The patient was edentulous. X-rays disclosed an approximately 3.0 cm radiolucent lesion surrounding the crown of an unerupted mandibular third molar tooth. It was thought to be a dentigerous cyst but upon removal it turned out to be a solid lesion. A diagnosis of fibrosarcoma was rendered by the local pathologist and the patient was referred to Truman Medical Center where a jaw resection and neck dissection was done. Your slide is of the mandibular tumor. Although slides are not included, three cervical lymph nodes contained tumor.

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ELLIS FISCHER STATE CANCER HOSPITAL
AND CANCER RESEARCH CENTER
ORAL PATHOLOGY SEMINAR #64
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CASE # 1 (78-15745)

MALIGNANT FIBROUS HISTIOCYTOMA
(Contributed by Albert M. Abrams, D.D.S., M.S.,
Professor of Pathology, Univ. of Southern
California School of Dentistry, Los Angeles, CA)

Dr. Berthrong from Colorado Springs and Dr. LeGal from Strasbourg consider the possibility of being metastatic. Drs. Yetter and Fay from the Wm. Beaumont Army Medical Center in Texas also added, "Rule out rhabdomyosarcoma and melanoma". Dr. Glass from Oklahoma and Drs. Dunlap and Barker from Kansas City also consider the possibility of a rhabdomyosarcoma. Drs. Tarpley and Corio from Bethesda, MD commented, "Malignant neoplasm - rule out metastatic giant cell carcinoma from lung - also considered were malignant fibrous histiocytoma and spindle cell carcinoma". Dr. Roger Terry, Chief of Surgical Pathology at LAC/USC Medical Center agreed with the diagnosis of malignant fibrous histiocytoma. Drs. Casas, Venditti and Davila from The Hospital Britanico, Buenos Aires, agree with the diagnosis of Fibroxanthosarcoma ? Rhabdomyosarcoma ?

CASE # 2 (04407/79)

AMELOBLASTOMA
(Contributed by Yvon LeGal, M.D., Institut
D'Anatomie Pathologique, Strasbourg, France)

This was also the diagnosis of Dr. Hori from West Virginia, Drs. Ackerman and Sciubba from Long Island, Drs. Dunlap and Barker from Kansas City. Dr. Glass and associates from Oklahoma commented, "Keratinizing, calcifying odontogenic cyst with an ameloblastoma arising in the wall". This was also impressions of Dr. Abrams from USC and Dr. Shafer from Indiana. Dr. Weathers from Emory made the following commentary: "We would consider this essentially as a mixed odontogenic tumor previously unclassified. We have seen two very similar cases; one from the Univ. of Southern California and one from the Univ. of Connecticut. I think this current case could very well be described as a peculiar odontogenic proliferation within a cyst or some might prefer to call this a Gorlin cyst with a peculiar epithelial proliferation. I do not think I would want to call this an ameloblastoma in association with a Gorlin cyst". Drs. Tarpley and Corio from Bethesda, MD commented, "Atypical Gorlin - areas of cellular pleomorphism make it difficult to predict the biologic behavior of the neoplasm". Drs. Fay and Yetter from Wm. Beaumont Army Medical Center diagnose, "Squamous cell carcinoma arising in a Gorlin cyst".

CASE # 3 (S79-349 2 D)

MUCORMYCOSIS
(Contributed by Henry A. Azar, M.D., Chief
Laboratory Service and Professor of Pathology,
Univ. of South Florida College of Medicine,
Tampa, Florida)

Without exception everyone interpreted as the result of fungal infection. To the question of why the orbital exenteration was done with the history that was given, Dr. Azar commented, "This 62 year old white male vacationing in Florida, known to have diabetes for 15 years, was first seen on 1/27/79 in a community hospital because of right orbital pain and right maxillary tenderness with a black patch on the right hard palate. This followed by a few days an extraction of a right bicuspid. Biopsy of the palate at the community hospital showed nonseptate hyphae. The patient was then referred to this hospital for the definitive care of the presumptive diagnosis of mucormycosis. Smears of the hard palate again,

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CASE # 3 Continued

showed broad nonseptate hyphae consistent with *Mucor* or *Rhizopus*. The patient was started on Amphotericin-B as well as energetic treatment of his diabetes. The next day, because of the rapidly progressive orbital cellulitis, weakness of extra-ocular muscles, proptosis and a "frozen" orbit, the patient underwent not only a total maxillectomy-ethmoidectomy, but also exenteration of the right eye. A debridement done on 1/31/79 showed residual necrotic tissue and bone with *Mucor* organisms.

In addition to identification of hyphae consistent with *Mucor*, culture on 2/7/79 yielded a *Mucor* species. The patient expired on 2/6/79. Autopsy revealed central nervous system edema together with extensive bilateral pneumonia.

This hospital has had prior experience with another case of mucormycosis. This individual is still living and well following exenteration and radical maxillectomy. He wears a prosthesis and his course has been uneventful except for having been mugged in downtown Tampa. This resulted in loss of his prosthesis which had to be replaced".

CASE # 4 (79-445)

EPIDERMOID CARCINOMA

(Contributed by Jose M. Hori, M.D., Davis Memorial Hospital, Elkins, W.V.)

Almost without exception this was the overwhelming diagnosis. Dr. Glass from Oklahoma commented, "I call it a high grade mucoepidermoid carcinoma". Dr. Shafer added, "Carcinoma, probably of ductal origin". Drs. Tarpley and Corio from Bethesda, MD commented, "Squamous cell carcinoma - the neoplasm has features of intraductal alveolar carcinoma ala shear". Dr. Ackerman from Long Island calls it carcinoma with basaloid features. Dr. Berthrong from Colorado Springs commented, "This would appear to be a very well differentiated but still invasive squamous cell carcinoma undermining the surface mucosa. I do not see the take off point. I cannot find mucin to suggest a muco-epidermoid tumor of salivary gland. But I suppose that should be looked for by special stains and more sections. I suppose that this could be the undermining portion of a verrucal carcinoma, but I do not see any evidence for that on the surface. I cannot find anything to suggest an adenoid cystic carcinoma. My diagnosis is squamous cell carcinoma". Dr Casas and associates from Buenos Aires called it epidermoid carcinoma, small cell variant.

CASE # 5 (S-79-15543, B.C.H.)

EPIDERMOID CARCINOMA

(Contributed by William C. Bucher, M.D., Boone County Hospital, Columbia, Missouri)

This was also the diagnosis of Drs. Ackerman and Sciubba from Long Island, Drs. Dunlap and Barker from Kansas City, and Dr. Wesley from Detroit. Dr. Shafer from Indiana commented, "A beautiful example of a Keinsasser tumor or, as he called it, a speichelgang-carcinome or salivary duct carcinoma. (Arch. Klin, Exp. Ohren Nasen Kehlkopfheilkd. 92:100, 1968)".

CASE # 6 (S-78-2076-A)

MELANOTIC NEUROECTODERMAL TUMOR OF INFANCY

(Contributed by J.B. Hanson and R.F. Graham, D.D.S., Oral Surgeons and A. Giuliani, D.O., Pathologist, Charles E. Still Hospital, Jefferson City, Missouri)

There was an unanimous agreement in this diagnosis.

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CASE # 7 (STIPE)

NERVE SHEATH NEOPLASM (BENIGN OR LOW GRADE
MALIGNANCY)(Contributed by Charles Dunlap, D.D.S., Univ.
of Missouri School of Dentistry, Kansas City, MO)

Dr. Spjut from Houston calls it well differentiated fibrosarcoma. This was also the diagnosis of Dr. Wesley from Detroit. Drs. Fay and Yetter from Wm. Beaumont Army Medical Center call it low grade fibrosarcoma, with xanthomatous changes. Dr. Abrams from USC commented, "I prefer fibrosarcoma for this tumor. Although large nerves are present. I see no good evidence of neural origin. I would also not call it fibrous histiocytoma. The foam cells are probably histiocytes and secondary to fat invasion and necrosis". Dr. Shafer from Indiana calls it low grade fibrosarcoma. Drs. Tarpley and Corio from Bethesda, MD call it "Atypical fibroxanthoma - rule out storage disease". Dr. Weathers from Emory stated, "Fibroxanthoma, however, the possibility of a lipid storage disease should be investigated". Drs. Scuibba and Ackerman from Long Island call it "Malignant Schwannoma. The identity of granular cells remains uncertain' perhaps they represent degenerating Schwann cell elements or maybe even histiocytic cells". Dr. Berthrong from Colorado Springs commented, "This interesting soft tissue neoplasm cannot be identified unequivocally by me without special stains and electron microscopy. The large foam cells that are scattered throughout at first I thought were granular cells and, therefore, I was going to make a diagnosis of malignant schwannoma forming granular cells. However, under oil immersion these appear to be vacuolated cells rather than granular so I suspect that they are foam cells. They are certainly striking and I cannot remember a soft tissue neoplasm with these present. If the neoplasm is destroying fat, however, macrophages could assume this appearance. I wish I could see a fat stain to be sure that they are foam cells rather than granular cells. My diagnosis is soft tissue neoplasm, ? malignant schwannoma, ? leiomyosarcoma". The following comments were submitted by Drs. Dunlap and Barker: "The biopsy revealed a proliferation of spindle shaped cells often exhibiting a whorled or herringbone pattern; however, no typical Verocay bodies were noted. Nuclei were oval or elongated having both blunted and pointed ends. The tissue was mainly of wavy fibrillar nature with some areas having a slightly more loose, almost myxoid appearance. Mitotic figures were not noted; however, occasional areas showing enlarged hyperchromatic, bizarre nuclei were encountered. The tumor was not encapsulated and involved neighboring fat tissue in a diffuse fashion. A trichrome stain ruled out a leiomyoma and we favored the diagnosis of a benign schwannoma, especially considering numerous neural-like elements within the tumor mass. Bodian stains however, were largely negative except for faintly positive areas such as this area shown. We could not completely determine that these areas were not pre-existent non-neoplastic nerve bundles. A neuropathologist was consulted who favored the diagnosis of an unusual benign schwannoma, unusual in that it was not well-delineated. It was decided to resect the mandible due to the possibility of low grade malignancy. At his time EM was performed. Cells occasionally showed well developed granular ER and prominent Golgi. Extracellular collagen was also occasionally abundant. No typical basement membranes of Schwann cells were identified. According to several investigators reporting at recent EM conferences, these findings do not completely rule out neural origin. The final diagnosis in this case may remain undecided since the patient died post-operatively of a pulmonary embolism. We still favor the diagnosis of a benign schwannoma; however, low-grade malignancy can not be completely ruled out. We can find only 39 cases of benign intrabony neural neoplasms of the jaws. Of these, the majority are neurofibromas, with only 3 of these having neurofibromatosis. We have 2 additional unpublished cases associated with Von Recklinhausen's disease. Intrabony nerve tumors are rare in any bone; however, the mandible appears to be the most common location in the entire skeleton. In the 3rd edition of Bone Tumors, Dahlin notes only 10 tumors in the entire skeleton with 4 located in the jaws, all within the mandible. Our search supports that the mandible is indeed the most commonly involved bone

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CASE # 7 Continued

with the posterior mandible being the usual location. Symptoms vary as noted with swelling and pain being the most common complaint. Many patients had both swelling and pain as their initial complaint. The tumors are more common in women with an average age of about 30. Local excision is the recommended treatment with recurrence reported in only 5 cases. Neurofibromas more commonly recur possibly related to the lack of a definite capsule. We know of 2 such cases which eventually required partial mandibular resection for complete removal".

CASE # 8 (S-1219-79)

UNDIFFERENTIATED MALIGNANT TUMOR - PROBABLY ANAPLASTIC CARCINOMA - PRIMARY (Contributed by Drs. Lin and Dodhia, Truman Medical Center, Kansas City, Missouri)

The diagnosis of carcinoma pleomorphic, N.O.S. was offered by Dr. LeGal from Strasbourg. Drs. Ackerman and Seiubba from Long Island wrote, "Undifferentiated malignant tumor. Rule out malignant melanoma. Electron microscopy would be of great help in this particular case". Dr. Spjut from Houston calls it carcinoma; malignant melanoma cannot be excluded. Drs. Tarpley and Corio from Bethesda, MD diagnosed; "Poorly differentiated malignant neoplasm melanoma vs. carcinoma". Dr. Abrams from USC wrote, "I strongly favor a diagnosis of melanoma. I suppose adenocarcinoma and rhabdomyosarcoma would have to be considered but the overall morphology and cellular characteristics are strongly suggestive of melanoma. I searched for pigment but could find none". The diagnosis of Dr. Shafer from Indiana is, "This is tough. Pleomorphic rhabdomyosarcoma vs. melanoma". Dr. Weathers from Emory wrote the following: "We thought that an epithelioid osteosarcoma was the best diagnosis of this lesion. A possibility of a metastatic deposit of melanoma from the head and neck area might also be considered, however, I feel more comfortable with the former diagnosis". Dr. Wesley from Detroit, Drs. Glass, Young, and Rohrer from Oklahoma call it alveolar rhabdomyosarcoma. Dr. Hori from West Virginia calls it malignant melanoma; this was also the diagnosis of Dr. Casas and associated from Buenos Aires.