Nevoid basal cell carcinoma (Gorlin) syndrome (NBCCS) is a well-recognized entity dating to at least early Egyptian time. One of the first documented cases was reported in 1894. Inheritance is autosomal dominant with an almost complete penetrance and variability in expression. Major clinical signs include multiple nevoid basal cell carcinomas, odontogenic keratocysts, skeletal anomalies and ectopic calcification.

 CASE REPORT
We treated a 15-year-old white male who had a lytic lesion in the left side of the mandible. A panoramic radiograph showed multiple mandibular cystic lesions and a cystic region in the left maxilla. The left mandibular and maxillary cysts appeared associated with impacted molars. A preoperative CT series permitted surgical removal of these cysts.

Family history. For this patient, there is a documented paternal history of this syndrome. An odontogenic keratocyst was successfully removed from the patient's anterior mandible 10 years ago. His sister, who also has NBCCS, has odontogenic keratocysts associated with impacted teeth.

**Abstract**

More than 100 anomalies are associated with this syndrome.

In this case, a cyst removed from a 15-year-old male was diagnosed as an odontogenic keratocyst. The long history of this syndrome with its associated problems is described.

Clinically the patient has a marfanoid appearance. He is 6 feet 7 inches tall with facial characteristics common to the syndrome—frontal bossing, hypertelorism and sunken eyes. The patient is under the care of a dermatologist for multiple basal cell nevi. Some nevi were removed and histopathologically confirmed (elsewhere) as basal cell carcinoma. The patient has palmar and plantar pits with associated dyskeratosis. Chest radiographs reveal rib bifurcation.

Surgery. In the operating room, general anesthesia was achieved with right nasal tracheal intubation. A mucoperiosteal flap in the mandibular right retromolar area was reflected. Exudate was observed, aspirated and sent for pathological evaluation of contents for keratin. The cyst was removed intact.

We found no perforation of cortical bone or neurovascular bundle. For the maxillary left retromolar area, a mucoperiosteal flap was reflected and perforation of the buccal cortex was observed. We found an impacted maxillary second molar superior and buccal to the root of the first molar. The impacted tooth and overlying bone were removed.

The bilobulated cystic lesion was removed intact from the left maxilla. It had perforated into the maxillary antrum. All bony fragments were removed and margins smoothed. The mucoperiosteal flap was repositioned and sutured. In the mandibular left molar region, the distally angled, partially impacted second molar was extracted. A mucoperiosteal flap was reflected. The cystic lesion was curetted from the buccal aspect, carried posteriorly and superiorly into the ascending ramus.

We found that the cystic lesion adhered to the tissues on the lingual aspect. The buccal cortex had a perforation about 1 centimeter in diameter. The impacted tooth was removed with the cystic lesion. After inspection, irrigation and
Curettage, the flap was repositioned. Estimated blood loss was 12 to 14 cc. There were no postoperative complications.

Radiology review. A panoramic radiograph of the lower facial bones and mandible shows sharply demarcated cysts within both mandibular rami (Figure 1). Within the left mandibular ramus, the cyst is 2.5 cm x 1.4 cm x 3.7 cm. The right mandibular cyst is about 2 cm x 1 cm x 2 cm. A 3 cm x 2.8 cm x 3 cm odontogenic cyst is also seen within the left maxillary sinus floor. There is no residual lesion apparent within the anterior mandible.

A coronal non-contrast CT scan shows both the keratocyst protruding into the maxillary sinus and premature falx calcification (Figure 2). Preoperative CT scanning provided a multiplanar view of all lesions and their relationship to nearby structures. For this case, CT was more useful than MRI to provide the fine bone detail.

Pathology review. A culture of the right retromolar exudate encountered during surgery was negative for bacterial growth. The content of the right mandibular cyst, submitted separately for evaluation, consisted only of keratinous debris. Three macroscopic and grossly intact cysts were evaluated, varying in diameter from 2 to 3.5 cm (Figure 3). The cyst walls varied in thickness, from 1 to 3 mm. Each contained thick, creamy, green-tan, sebaceous-type contents. No other sections of bone or tissue showed growth.

Microscopic examination of the maxillary and mandibular cysts showed features of odontogenic keratocyst. These included the presence of a thin keratinizing stratified squamous epithelial lining of six to eight cell layers, a prominent basal cell layer without the formation of rete ridges and a thin fibrous tissue wall (Figure 4). Foci of chronic inflammation and fibrosis were also noted. There was no evidence of associated squamous cell carcinoma or ameloblastoma. Microscopic sections of soft tissue from the left maxillary tuberosity showed an incipient “daughter” keratocyst 1 mm in diameter (Figure 4, insert).

LITERATURE REVIEW
There are more than 100 anomalies associated with NBCCS. In this paper, anomalies are divided into subgroups.
Skeletal. About 75 percent of these patients have skeletal anomalies. Bifurcated, anteriorly splayed, fused, partially missing, hypoplastic and cervical ribs have been noted. Bifurcation is the most common rib anomaly, and is often seen in more than one rib. Vertebral anomalies in the upper thoracic area include spina bifida occulta and scoliosis. Bridging of the sella turcica and synostosis has been seen. Cervical and/or thoracic vertebral fusion has been documented. Frontal and biparietal bossing results in a head circumference of 60 centimeters or more in 25 percent of patients.

Patients are often tall with a marfanoid build. Apparently, there is a correlation between head circumference and height. NBCCS has often been considered as a "macrocephaly syndrome." Sinuses are hyperpneumatized in 60 percent of cases because of the absence of intranasal septa. Well-developed supraorbital ridges give eyes a sunken appearance. Small pseudocystic-lytic bone lesions have occurred in the phalanges, metapodial, carpal and tarsal bones, long bones, pelvis and calvaria.

Dentition. Odontogenic keratocysts have occurred in 65 to 100 percent of patients with this syndrome. These cysts are three times more common in the mandible, and can be single, isolated, multiple, bilateral, unilocular or multilocular and asymmetric. Cysts can cross the midline in both the maxilla and the mandible. As a result of tension from the cyst content, jaw swelling, dull pain and intraoral drainage of contents are common. There is always the potential for these cysts to transform to ameloblastoma from the usual basal cell carcinomas in that in NBCCS, the tumors usually appear between puberty and age 35. There can be literally hundreds and thousands of lesions. Lesions are pink-pale brown papules, 1 to 10 mm in diameter. They are common on the face, neck and upper trunk. But the periorbital area, eyelids, nose, upper lip and malar area are most often affected.

These lesions, unlike the common basal cell carcinomas, occur in both sun-exposed and unexposed areas, but are more frequent in exposed areas. Palmar and plantar pits, with associated dyskeratosis, which may even be pathognomonic, are extremely common, and have been observed in 50 to 75 percent of patients. There have been several documented cases of basal cell carcinoma originating in the base of these pits.

Ophthalmologic. Internal strabismus and congenital blindness caused by cataracts have been described. A broad nasal root is frequently seen, which may be associated with true ocular hypertelorism.

Facial appearance. The face is characterized by...
hypertelorism with frontal bossing and broad nasal root.\textsuperscript{9,10} Frontal and temporoparietal bossing with well-developed supraorbital ridges give eyes a characteristic sunken appearance.\textsuperscript{9,12} Eyebrows are often heavy and fused.\textsuperscript{4} There is often an increase in the size of the cranium with an occipito-frontal circumference of at least 60 cm.\textsuperscript{4} Patients can also have less than normal amounts of facial hair and diminished perspiration.\textsuperscript{4}

Central nervous system. Mild mental retardation (in about 3 percent) and schizophrenia have been reported in these patients.\textsuperscript{4,9,10} Agenesis of the corpus callosum and congenital hydrocephalus is apparent.\textsuperscript{9,10,14} Calcification of the falx cerebri is seen in 85 percent of patients, as compared with 5 percent in the normal population.\textsuperscript{4} Calcification of the tentorium cerebelli, petroclinoid ligament, dura, pia and choroid plexus has been observed.\textsuperscript{4} There has been an association of NBCCS with a high incidence of medulloblastoma. But a definite correlation cannot be made since this occurs within the first two years of life, before the usual symptoms of NBCCS are apparent.\textsuperscript{4,10} Meningioma and craniopharyngioma have also been described.\textsuperscript{4}

**SUMMARY**

Although NBCCS has an autosomal dominant pattern, 30 to 60 percent of patients do not have a positive family history.\textsuperscript{4,15} There is a correlation between older paternal age and new mutations.\textsuperscript{10} There appears to be an inherited predisposition to malignancy in these patients.\textsuperscript{1,11,14,17} Patients should have dermatologic examinations every three to six months. Nevi showing growth, ulceration or hemorrhage should be removed.

Although surgical removal is often indicated, recent studies suggest oral retinoid may be helpful.\textsuperscript{4,18} Odontogenic keratocysts need to be surgically removed. Several techniques, such as marsupialization and enucleation, followed by either primary closure or packing of the wound, have been used.\textsuperscript{18} Some authors feel that recurrence is rarer if the cyst is enucleated intact.

Patients should be aware of the high recurrence rate of these cysts.\textsuperscript{4,20} An annual clinical and radiologic oral surgery evaluation is advised. NBCCS is a multisystem disease,\textsuperscript{4} and the clinical treatment of patients involves many medical and dental specialties. A team approach is essential for patients’ health and well-being.

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