Housecalls

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INSIDE THIS ISSUE

EMERGING NEWS

Guidelines for Prescribing Opioids

The opioid epidemic and its associated overdose deaths have been widely reported in the past few years. In response, the Center for Disease Control issued a guideline for prescribing opioids for chronic pain (excluding cancer, palliative and terminal pain) in March 2016.

The guidelines were developed to reduce inappropriate prescribing practices with a goal of reducing opioid use disorder, overdoses and death. One focus was reducing the dosage and the duration of opioid treatment to the minimum necessary to relieve pain. It suggested that three days of treatment is often sufficient for the relief of pain, with more than seven days rarely being needed.

The guidelines encouraged the discussion of the dangers of overdose between physicians and patients on long-term, high-dose treatments. The question arises, are there signs of benefit?

There is an unavoidable lag time to collect data on prescriptions written and reported deaths due to overdose. However, preliminary data is encouraging. The prescribing of opioids peaked in 2012 with a rate of 81.3 opioid prescriptions per 100 persons in the US. The prescription rate in 2017 was down to 58.7 per 100, the lowest in a decade. There are still pockets of very high prescribing rates in the US with 16% of counties reporting enough prescriptions for every person to have had one.

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The Art & Science of Risk

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CASE #1

Hereditary Multiple Exostoses

By Regina Rosace, MD
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A 36-year-old male applies for life insurance. He states that he is in good health except for a history of Hereditary Multiple Exostoses. He sees an orthopedic surgeon yearly. His mother also has this diagnosis and is alive and well. He has had two surgeries for removal of benign lesions in his knee and elbow.

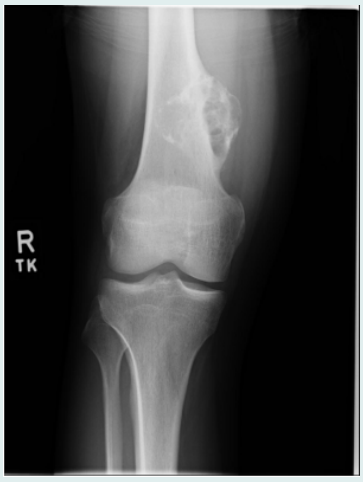
Definition

Hereditary Multiple Exostoses (HME) is an autosomal dominant condition characterized by abnormal growth of long bones mainly affecting the epiphyses. Its many synonyms include Multiple Osteochondromas (MO), Multiple Hereditary Exostoses (MHE), EXT, Multiple Hereditary Osteochondromas (MHO) and Multiple Cartilaginous Exostoses. Osteochondral growths (exostoses or osteochondromas), consisting of bone surrounded by a cap of cartilage, occur on the periphery of long bones.

The patient must have two or more exostoses on the appendicular or axial skeleton to fulfill the definition of HME. Approximately 80% of individuals with this condition have a positive family history. The major complaints of these patients are cosmetic deformities, pain and difficulty moving the joints. Osteochondromas are benign lesions that do not affect life expectancy. Rarely, these exostoses may transform into chondrosarcomas.

Presentation

The presentation often occurs in the second decade of life. Often it is an incidental finding, with males more than females. Incidence is estimated to be 1:50,000. The most common location is the around the knee or the proximal humerus. The osteochondroma can present as a painless mass or a painful mass with minimal trauma. The lesions can cause pain, decreased range of motion, deformity or pathologic fracture. The growth plates can be affected, with resultant short stature or angular deformities. Fifty percent of patients present with a clinically visible tumor by age 5, 80% by age 10. The average number of exostoses per patient is six.



Osteochondroma of distal femur accessed August 13, 2018 from Bing images, free to share and use

Imaging

The best form of imaging for this is plain radiographs in at least two planes.

Genetics

HME is an autosomal dominant condition with 95% penetrance. Two EXT genes account for 90% of the lesions. EXT1 is located on the distal end of chromosome 8, and EXT2 is located on chromosome 11. EXT1 and EXT2 have been associated with both familial hereditary exostoses and spontaneous multiple hereditary exostoses.

Mutations in either of these genes result in clinically indistinguishable exostoses. The proteins resulting from EXT1 and EXT2 are glycosyltransferases, which catalyse heparin sulphate polymerization and appear to be a complementary pair that form a stable enzyme complex in vivo. Research has suggested that the EXT genes function as tumor suppressor genes.

Treatment

Treatment is generally supportive. Surgery is required for the management of pain and deformation. Also, monitoring is necessary for the possibility of malignant transformation of lesions to secondary chondrosarcomas. This occurs in approximately 0.5% to 5% of lesions, so yearly radiographs are advised. The five-year survival of a chondrosarcoma is 90%. A wide surgical excision is the treatment of choice.

Prognosis

Chondrosarcomas are benign and do not affect the life span of an individual. There is a low incidence (0.5% - 5%) of transformation to malignant lesions. These lesions have a five-year survival rate of 25-90%, depending upon location.

Malignant transformation occurs most often in osteochondromas of the spine, scapula, pelvis and proximal femur. Benign lesions generally have cartilage caps < 2 cm thick in adults and tend to stop enlarging when the bony growth plates close.

Returning to the Case

This gentleman has a history of HME and has yearly follow ups with his orthopedic surgeon. His mortality risk appears to be low to near standard.

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CASE #2

Plummer-Vinson Syndrome

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A 34-year-old woman applied for life insurance. She was 5 feet 2 inches tall and weighed 230 lbs. Review of the medical history revealed thyroidectomy and hysterectomy (for endometriosis) in the distant past. Laboratory abnormalities included hemoglobin of 10.6 to 11.8 g/dl in 2017 and 2018.

There was an elevated erythrocyte sedimentation rate (ESR) of 61 and 69 mm/hr in 2017. There was a remote history of pancreatitis, cholecystectomy and a biliary stent. There was also a history of fibromyalgia. The proposed insured was prescribed thyroid replacement and iron supplementation. In addition, there was a diagnosis of iron deficiency anemia due to sideropenic dysphagia without details.

What is sideropenic dysphagia and what are the mortality implications?

Sideropenic dysphagia is more commonly known as Plummer-Vinson syndrome (PVS) in the United States. The disorder is also known as Patterson-Brown-Kelly syndrome in the UK.

It is a rare disorder consisting of the clinical triad of dysphagia, an esophageal web or membrane and iron deficiency anemia. Other findings, probably related to the iron deficiency, can include glossitis, cheilosis and koilonychia (spoon-shaped nails).

Incidence

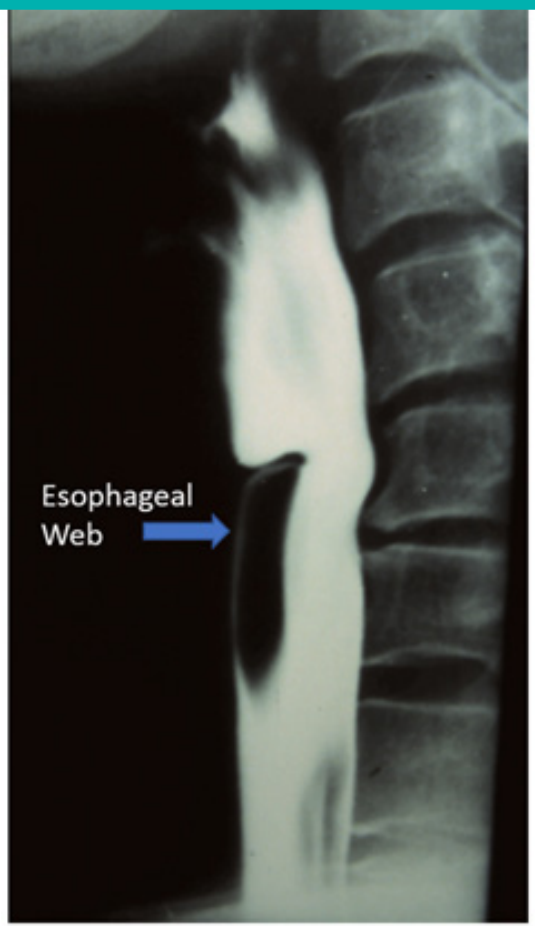
While exact figures on the frequency of PVS are unknown, it was not uncommon in Scandinavian countries in the early 1900's. More recently it is extremely rare, occurring primarily in Caucasian women in their fourth to seventh decades of life. Very rarely it can present in children, adolescents or men.

The exact pathogenesis is unknown, but is thought to be related to a combination of autoimmune, genetic and dietary influences. An association with celiac disease, thyroid disease and rheumatoid arthritis has been noted.

Reduction in the incidence of PVS over time is thought to be related to better nutrition and less iron deficiency in the population. However, in areas of Africa where malnutrition and iron deficiency anemia are endemic, PVS is very rarely encountered, raising the possibility that there must be other factors in the pathogenesis.

Dysphagia, often with solid foods, is the most common presenting complaint. It can be intermittent, progressive and associated with weight loss. Esophageal webs can be identified using barium swallow, videoflouroscopy or upper gastrointestinal endoscopy. The webs are usually below the cricoid cartilage and adhere to the anterior wall of the esophagous. Once other causes of dysphagia and iron deficiency anemia are excluded, the diagnosis can be made based on the classic triad.

FIGURE 1 —ESOPHAGEAL WEB ON BARIUM SWALLOW



Treatment

Treatment of PVS consists of iron replacement or other treatment of anemia. Often symptoms of dysphagia will improve with resolution of the anemia. If symptomatic, dysphagia can be treated with balloon dilatation. PVS is recognized as a risk factor for developing pharyngeal or esophageal squamous cell carcinoma. For that reason, some experts recommend monitoring with periodic endoscopy.

Returning to the Case

There were no details as to how the diagnosis was made. It would be prudent to get details of the work-up and the monitoring given the association of PVS with squamous carcinoma. In general, those with PVS respond well to iron treatment and esophageal dilatation as needed. If a thorough evaluation was done and ongoing monitoring is planned, there would be minimal excess mortality anticipated.

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EMERGING NEWS

Guidelines for Prescribing Opioids

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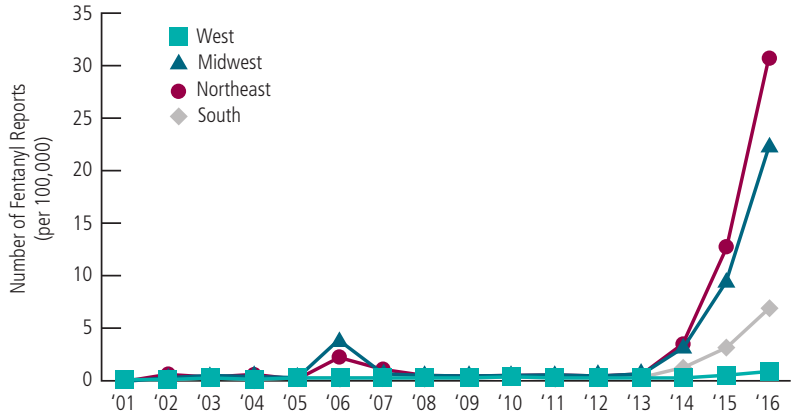
FIGURE 1 – REGIONAL TRENDS IN FENTANYL REPORTED PER 100,000 PERSONS AGED 15 OR OLDER, JANUARY 2001-DECEMBER 2016



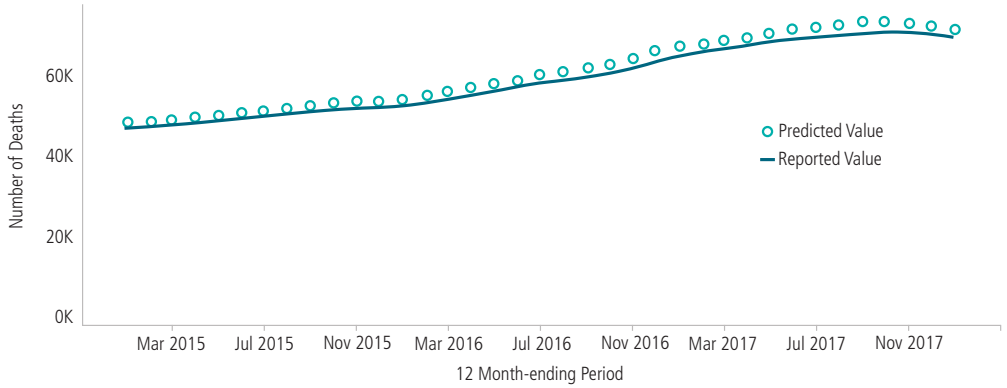
Note: US census 2016 population data by age were not available for this publication. Population data for 2016 were imputed.

The Impact of Fentanyl

Overdose death rates appear to be linked to prescription opioid exposure and appear to be related to the availability of illicit fentanyl. Fentanyl is a synthetic opioid that is approximately 100 times more potent than morphine and 50 times more potent than heroin. Illegal fentanyl is often mixed into heroin or pressed into pill forms that mimic prescribed opioids. Slight miscalculations in illicit formulation can lead to overdose and death.

Unfortunately, the number of fentanyl encounters collected by the DEA has increased dramatically (Figure 1). Interdiction of the flow of illicit fentanyl, treatment of opioid abuse disorder and awareness of the dangers of using illicit opioids are all important components of a solution to the problem of drug overdose deaths. The CDC monitors overdose deaths, and there may be the early beginnings of leveling off or improvement as shown in Figure 2.

FIGURE 2 – 12-MONTH ENDING PROVISIONAL COUNTS OF DRUG OVERDOSE DEATHS IN THE US



12-month ending provisional counts of drug overdose deaths in the US based on data available for analysis on August 5, 2018.

Another encouraging opioid trend can be found in the Drug Testing Index™, a report on workforce drug positivity produced by Quest Diagnostics. Quest summarizes the prior year test results for their customers doing work-related employee drug testing. They found that the opiate positivity rate in the general US workforce declined 17% from 2016 to 2017. Urine drug testing for heroin revealed a decline of 11% between 2016 and 2017 and hit a three-year low.

But the news from employee and pre-employee drug testing was not all good. The rate of positive urine cocaine tests had increased 7% in 2017 over 2016, marking the fifth year of consecutive increases. Methamphetamine positivity also increased dramatically in certain areas of the country. Overall the combined drug positivity rate for the US workforce held steady year over year at 4.2% of specimens testing positive.

It does appear that efforts toward reversing opioid prescribing are beginning to have a positive effect, but continued programs to prevent overdose are needed. And ongoing monitoring of the situation is required.

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SOLVE

ECG Puzzler...

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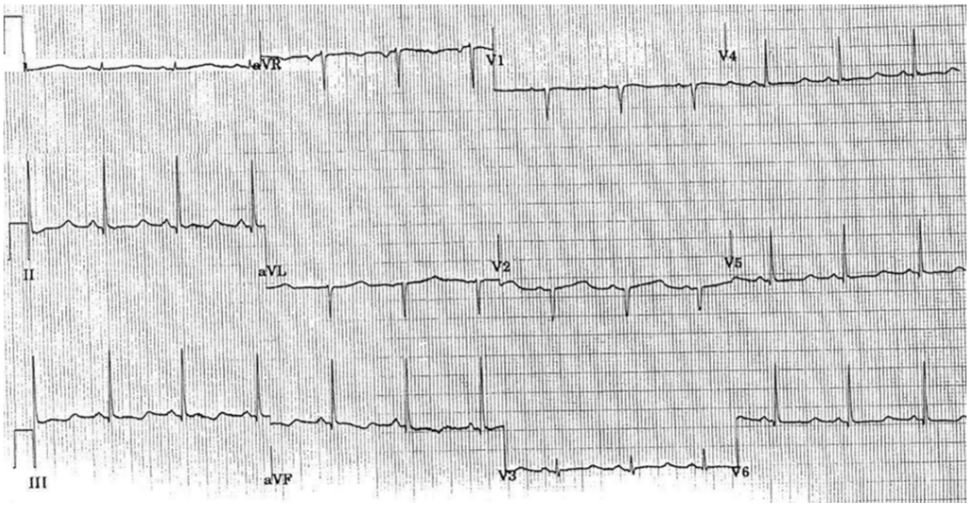

Here is the latest ECG Puzzler to solve.

A 43-year-old woman underwent thyroidectomy for a follicular cancer of thyroid. A few days later, she complained of muscle cramps in the back and sharp chest pain. She had no cardiovascular risk factors and no family history of sudden death. Her physical, BMI and vitals were within normal limits. An ECG was obtained. What does it reveal?

To find the answer, visit the Housecalls page at www.scorgloballifeamericas.com. Click on September 2018 Puzzler to confirm your findings.

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