by the presence of proximal and distal polyphasias, many complex, in all muscles tested. From these results, as well as the previous studies, it was concluded that the origin of her neuropathy had been primarily peripheral demyelination changes with axonopathic features.

She continued to improve and was discharged after the 57th hospital day. Over one year later, she is without recurrence of her neurologic problem.

**DISCUSSION**

The Ad Hoc Committee sponsored by the National Institute of Neurological and Communicative Disorders and Stroke (NINCDS) divided the diagnostic features of GBS into two categories—those required for the diagnosis and those strongly supportive of the diagnosis. The only features required for diagnosis are progressive motor weakness in more than one limb and areflexia. Unfortunately, these features may be masked in the critically ill by prolonged sedation, paralysis, and ventilatory dependence. Once other etiologies have been ruled out, then supportive diagnostic features may be used. Classically, these are elevation in CSF protein with CSF mononuclear cell count less than 10 per ml, NCV slowing, and increase in distal latencies. In the absence of clinical signs and equivocal studies of CSF, EMG, and NCV, the diagnosis relies on an understanding of the variant features with which GBS can present. Among the variants pertinent to this discussion are the lack of a rise in CSF protein in the period of one to ten weeks after the onset of symptoms (though rare) and entirely normal NCS (an incidence of 20 percent). Other investigators report that patients with GBS who have normal peripheral NCV with severe paralysis may have segmental demyelination in the proximal portion of nerve roots and that conduction may be only mildly slowed in patients in whom axonal degeneration occurs. In fact, in studies to date, axonal degeneration is the one indication of prolonged disability that EPS can demonstrate.  

Critical illness polyneuropathy as it has been defined by Zochodne et al, is an entity found only in patients with sepsis and multiorgan failure which is differentiated from GBS in studies of the CSF and electrophysiology. The CIP demonstrates normal or mild CSF protein elevation and primarily axonal degeneration. These, according to the standard definition, can be considered variant features of GBS. The etiology of CIP is unclear, but the condition only seems to resolve as sepsis and multiorgan failure improve. No specific therapy for this condition exists.

Finally, this case is the second GBS in a severe asthmatic in whom the diagnosis relied on variant presentations of CSF and/or electrophysiologic studies which do not always readily distinguish it from CIP. If CIP has an autoimmune origin, as is thought GBS does, then plasmapheresis may prove beneficial in its therapy as well. Familiarity with the subtleties of both diseases is essential in aiding the intensivist in making diagnostic and therapeutic decisions.

**REFERENCES**


**Hypercalcemia in Atypical Bronchial Carcinoid Tumors**

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Atypical carcinoid tumors of the bronchial tree are uncommon. Their tendency to metastasize is well recognized, characteristically producing osteoblastic bone deposits without disturbance of calcium homeostasis. We report two patients who presented with hypercalcemia and osteolytic bone metastases following surgical removal of atypical bronchial carcinoid tumors. In one of the patients, chemotherapy induced remission and controlled the hypercalcemia.

(Chest 1989; 96:1206-08)

Carcinoid tumors represent about 2 percent of all pulmonary neoplasms. The cell of origin is thought to be the Kulchitzky cell of the AFUD (amine precursor uptake and decarboxylation) series, similar to that of the highly malignant small cell lung cancer (SCLC). It has been suggested that carcinoid and small cell tumors are the two ends of a spectrum of malignant change affecting the AFUD cell line. Intermediate in metastatic potential between these is the atypical carcinoid.

Although initially described in 1944, the histologic characteristics of atypical carcinoid tumors were not fully documented until 1972. Such tumors are uncommon, accounting for approximately 10 percent of bronchial carcinoid tumors. Roentgenographically a solitary mass, possibly lobulated, lying in an upper lobe may be seen.

Metastases in bones are uncommon in typical carcinoid tumors originating from the bronchus, but when found, are almost always osteoblastic, the presence of lytic secondaries being more suggestive of a primary site in the gastrointestinal tract. Hypercalcemia is rare in typical bronchial carcinoid tumors and appears undocumented in atypical bronchial carcinoid tumors.

We report two patients with atypical carcinoids who presented with hypercalcemia and osteolytic bone secondaries following resection of the primary lung tumor.

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CASE REPORTS

CASE 1

A 50-year-old man who smoked 80 cigarettes per day presented with a ten-month history of chest pain and cough. Physical examination was normal but a 35 mm round mass was evident on his chest x-ray film. Bronchoscopy showed tumor in the left lower lobe, biopsy of which showed an atypical carcinoid tumor. No metastases were detected clinically and he underwent left lower lobectomy, histology confirming the diagnosis of atypical carcinoid (Fig 1 and 2), with hilar node involvement (stage T2 N1).

He presented three months later with a three-day history of malaise, thirst, backache, facial paraesthesia, thigh pain, and weakness of both legs. On examination, he had an enlarged firm irregular liver, reduced sensation over the first and second divisions of the right trigeminal nerve with loss of the corneal reflex. Signs of a spastic paraparesis were found in his lower limbs. Abdominal ultrasound confirmed multiple hepatic metastases and films of his spine showed a compression fracture at T12 with a total block at this site on myelography. Plasma calcium (corrected to an albumin of 40 g/L) had risen from 2.26 on the previous admission to 2.70 mmol/L (normal range 2.12 to 2.62 mmol/L). Palliative radiotherapy in a dose of 17.5 Gy split as two fractions was given to the spine, but he died one week later.

CASE 2

A 65-year-old woman who never smoked presented with a six-month history of unproductive cough and breathlessness for one month. On physical examination, she had signs of left upper lobe collapse which was confirmed on chest roentgenogram. At bronchoscopy, tumor blocked the left upper lobe. Biopsy specimens were unhelpful. Mediastinoscopy was normal so pneumonectomy was performed. Histology confirmed the tumor as an atypical carcinoid (Fig 3) with hilar node involvement (stage T2 N1).

She presented six months later with malaise, anorexia, nausea, constipation, and chest wall pain. On examination, she was dehydrated with a scalp metastasis evident. Osteolytic deposits in the ribs and thoracic spine were seen on the chest x-ray film, corresponding to her pain and an isotope bone scan showed numerous "hot spots." Her corrected plasma calcium was grossly elevated at 3.84 mmol/L.

Vigorous rehydration with intravenous saline solution plus diuretics altered neither the hypercalcemia nor her symptoms. Plicamycin (2 mg over 48 hours) caused a transient fall in the plasma calcium, but was associated with thrombocytopenia (Fig 4). In view of the widespread metastases, intravenous chemotherapy was

FIGURE 1. Atypical carcinoid tumor from case 1 showing prominent rosette and gland formation with destruction of bronchial cartilage (hematoxylin-eosin, original magnification ×160).

FIGURE 2. Atypical carcinoid tumor from case 1 at higher magnification showing rosette formation, mitotic activity and necrosis (hematoxylin-eosin, original magnification ×320).

FIGURE 3. Section of pneumonectomy specimen from case 2 showing an atypical carcinoid tumor with a solid pattern. Packeted spindle cells are separated by a fine fibrovascular stroma, mitoses are moderate in number but necrosis is absent (hematoxylin-eosin, original magnification ×160).

FIGURE 4. Changes in plasma calcium with treatment for case 2 following readmission.

PLASMA TOTAL Ca²⁺ (corrected), mmol/l

Days Following Readmission With
Hypercalcaemia
started, using the SCLC protocol ACE (doxorubicin, 50 mg per m² day 1; cyclophosphamide, 1 g per m² day 1; and etoposide, 120 mg per m² days 1-7; repeated every three weeks). She completed six cycles with an excellent clinical response, remaining well nine months after re-presenting. The scalp metastasis was no longer palpable and the plasma calcium which was beginning to rise after stopping the plicamycin remained within the normal range (Fig 4).

**Discussion**

The management and prognosis of atypical bronchial carcinoid tumors is not well established, due in part to the relative rarity of the tumor. Diagnosis is usually made at resection, when almost half (range 28 to 70 percent) of the patients will have peribronchial or hilar metastases. Despite this, intrathoracic tumor recurrence following resection is uncommon and lobectomy appears the correct surgical approach. Following surgery alone, between one third and one half of patients die over a period of 21 to 27 months. In one study in which details are given, six out of eight patients had evidence of metastases at diagnosis (extrathoracic in three), and four died within 21 months despite “curative” surgery. The role of adjuvant therapy is unknown, but in seven patients who received chemotherapy and/or radiotherapy after surgery, all were alive after a 23 to 127 month follow-up period. In a subsequent study, three patients who received adjuvant chemotherapy were alive and well between eight and 101 months with only one having evidence of tumor recurrence.

We describe two patients who presented with atypical bronchial carcinoid tumors and hilar metastasis who represented the wide spectrum of clinical behavior with hypercalcemia in six months. Although both patients had osteolytic metastases, we do not know whether the hypercalcemia was due to the bony metastases or to one of the other tumor-associated causes. Chemotherapy was used in the second patient in view of the widespread disease and our choice of drugs was based on the assumption that atypical bronchial carcinoids may be biologically related to small cell lung cancer and may behave in a similar fashion. This treatment was very effective with symptomatic improvement, progression of the scalp metastasis, reduction of the serum calcium which was beginning to rise after the plicamycin, and clinically disease-free survival up to nine months.

Until a large series objectively studying therapy is published, treatment of atypical bronchial carcinoid tumors, especially those which have disseminated, will remain empirical. Chemotherapy as first line treatment of atypical bronchial carcinoid tumors is unsuccessful, but appears the treatment of choice in patients with widespread disease, and since dissemination occurs after apparently curative resection, adjuvant therapy may well be indicated following surgery.

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**Hypertrophic Osteoarthropathy Associated with Pneumocystis carinii Pneumonia in AIDS**

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Hypertrophic osteoarthropathy (HOA) is a systemic disorder primarily affecting the bones, joints, and soft tissues and developing in association with another disease process. Acute pyogenic pulmonary processes (empyema, lung abscess) are occasionally accompanied by transient HOA, but reversible HOA has not previously been reported in the setting of PCP in AIDS. (Chest 1989; 96:1206-09)

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**Case Report**

A 37-year-old man was admitted with a one-month history of...