TABLE 389-2 CLINICAL MANIFESTATIONS OF RELAPSING POLYCHONDRITIS

Clinical Feature	Presenting	Cumulative
	Frequency, %	
Auricular chondritis	43	89
Arthritis	32	72
Nasal chondritis	21	61
Ocular inflammation	18	59
Laryngotracheal symptoms	23	55
Reduced hearing	7	40
Saddle nose deformity	11	25
Cutaneous	4	25
Laryngotracheal stricture	15	23
Vasculitis	2	14
Elevated creatinine	7	13
Aortic or mitral regurgitation	0	12

Source: Modified from PD Kent et al: Curr Opin Rheumatol 16:56, 2004.

patients, and CD4+ T cells have been observed at sites of cartilage inflammation.

CLINICAL MANIFESTATIONS

The onset of relapsing polychondritis is frequently abrupt, with the appearance of one or two sites of cartilaginous inflammation. The pattern of cartilaginous involvement and the frequency of episodes vary widely among patients. Noncartilaginous presentations may also occur. Systemic inflammatory features such as fever, fatigue, and weight loss occur and may precede the clinical signs of relapsing polychondritis by several weeks. Relapsing polychondritis may go unrecognized for several months or even years in patients who only initially manifest intermittent joint pain and/or swelling, or who have unexplained eye inflammation, hearing loss, valvular heart disease, or pulmonary symptoms.

Auricular chondritis is the most frequent presenting manifestation of relapsing polychondritis, occurring in 40% of patients and eventually affecting about 85% of patients (Table 389-2). One or both ears are involved, either sequentially or simultaneously. Patients experience the sudden onset of pain, tenderness, and swelling of the cartilaginous portion of the ear (Fig. 389-1). This typically involves the pinna of the ears, sparing the earlobes because they do not contain cartilage. The overlying skin has a beefy red or violaceous color. Prolonged or recurrent episodes lead to cartilage destruction and result in a flabby or

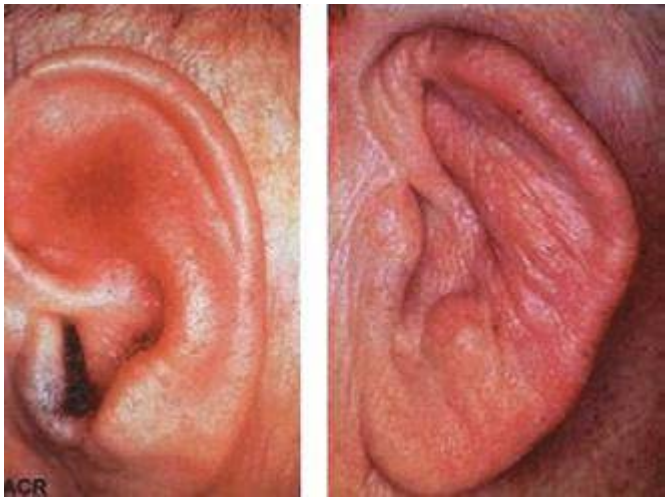

FIGURE 389-1 ***Left.*** The pinna is erythematous, swollen, and tender. Not shown is the ear lobule that is spared as there is no underlying cartilage. ***Right.*** The pinna is thickened and deformed. The destruction of the underlying cartilage results in a floppy ear. *(Reprinted from the Clinical Slide Collection on the Rheumatic Diseases, ©1991, 1995, 1997, 1998, 1999. Used by permission of the American College of Rheumatology.)*

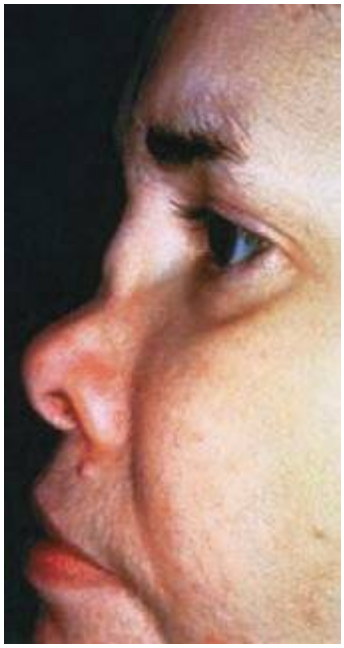

FIGURE 389-2 Saddle nose results from destruction and collapse of the nasal cartilage. *(Reprinted from the Clinical Slide Collection on the Rheumatic Diseases, ©1991, 1995, 1997, 1998, 1999. Used by permission of the American College of Rheumatology.)*

droopy ear. Swelling may close off the eustachian tube or the external auditory meatus, either of which can impair hearing. Inflammation of the internal auditory artery or its cochlear branch produces hearing loss, vertigo, ataxia, nausea, and vomiting. Vertigo is almost always accompanied by hearing loss.

Approximately 61% of patients will develop nasal involvement, with 21% having this at the time of presentation. Patients may experience nasal stuffiness, rhinorrhea, and epistaxis. The bridge of the nose and surrounding tissue become red, swollen, and tender and may collapse, producing a saddle nose deformity (Fig. 389-2). In some patients, nasal deformity develops insidiously without overt inflammation. Saddle nose is observed more frequently in younger patients, especially in women.

Joint involvement is the presenting manifestation in relapsing polychondritis in approximately one-third of patients and may be present for several months before other features appear. Eventually, more than one-half of the patients will have arthralgias or arthritis. The arthritis is usually asymmetric and oligo- or polyarticular, and it involves both large and small peripheral joints. An episode of arthritis lasts from a few days to several weeks and resolves spontaneously without joint erosion or deformity. Attacks of arthritis may not be temporally related to other manifestations of relapsing polychondritis. Joint fluid has been reported to be noninflammatory. In addition to peripheral joints, inflammation may involve the costochondral, sternomanubrial, and sternoclavicular cartilages. Destruction of these cartilages may result in a pectus excavatum deformity or even a flail anterior chest wall.

Eye manifestations occur in more than one-half of patients and include conjunctivitis, episcleritis, scleritis, iritis, uveitis, and keratitis. Ocular inflammation can be severe and visually threatening. Other manifestations include eyelid and periorbital edema, proptosis, optic neuritis, extraocular muscle palsies, retinal vasculitis, and renal vein occlusion.

Laryngotracheobronchial involvement occurs in ~50% of patients and is among the most serious manifestations of relapsing polychondritis. Symptoms include hoarseness, a nonproductive cough, and tenderness over the larynx and proximal trachea. Mucosal edema, strictures, and/or collapse of laryngeal or tracheal cartilage may cause stridor and life-threatening airway obstruction necessitating tracheostomy. Involvement can extend into the lower airways resulting in tracheobronchomalacia. Collapse of cartilage in bronchi leads to pneumonia and, when extensive, to respiratory insufficiency.

Cardiac valvular regurgitation occurs in about 5–10% of patients and is due to progressive dilation of the valvular ring or to destruction