

7.3 Natural history of idiopathic chronic pancreatitis

G. G. Tsiotos

Introduction

In the past, chronic pancreatitis (CP) was considered to be almost exclusively associated with overconsumption of alcohol (alcoholic chronic pancreatitis – ACP). During the past two decades idiopathic chronic pancreatitis (ICP) was recognized as a new entity. This was identified as CP with all its classical features, which was not associated to any alcohol intake. Thus, until very recently, CP was considered to be either ACP in about 70% to 90% of the time, or ICP in the remaining 10–30%. However, this convenient scheme has been challenged based on last decade's findings.

Until the early 1990s, all patients with non-ACP were automatically labeled as having ICP. Within this group however, there have been recognized several other conditions predisposing to or causing CP. Among them, more common and conceptually more important is the cystic fibrosis gene mutation (CFTR mutation – Cystic Fibrosis Transmembrane Regulator gene), which is now recognized in 25–48% of patients who were previously considered as having ICP [1–5]. About 15% of patients with “ICP” are now recognized as suffering from sphincter of Oddi dysfunction – type 1 (SOD-1). Another 5% have autoimmune CP, 1–5% hereditary CP [6], and about 1% pancreas divisum. Thus, from the initial 10–30% of patients with “ICP”, there remains only 25–30% of these patients with *true ICP*. Consequently, from the whole group of patients with CP, *only 3–9% are recognized today as truly having ICP*.

In addition to some very crucial terminology problems, which seriously affect the understanding of both ACP and ICP (see later), there have been yet other, even more exciting new epidemiologic findings, which altogether put the understanding of ICP and, for that matter, of CP as a whole into a totally new perspective [23–25]. Based on modern analysis, *it now seems that CP may be just two distinct forms of ICP (early-onset ICP and late-onset ICP), and the expression of one of them (late-onset ICP) depends a lot on environmental factors, such as alcohol intake or, to a lesser extent, smoking*. Thus, a deep and thorough understanding of ICP is of paramount importance for the appropriate, modern understanding of CP.

Terminology

The term “idiopathic chronic pancreatitis” was first introduced in the literature in 1976, when Rudolph Ammann identified a distinct group of older patients with all the clinical, biochemical, and imaging features of CP, with no history of alcohol intake whatsoever; for this subgroup he coined the term “senile chronic pancreatitis” [20]. After the introduction of this revolutionary concept, the pancreatic literature and the understanding of ACP and ICP has been hampered by one very important factor: *the lack of a universally accepted level of daily alcohol intake above which patients are considered as having ACP*. This issue, al-